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Predictors of Retrograde Aortic Arch Obstruction After Hybrid Palliation of Hypoplastic Left Heart Syndrome

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Abstract

A potential complication after hybrid stage 1 palliation for hypoplastic left heart syndrome (HLHS) is retrograde aortic arch obstruction (RAAO). This can lead to increased morbidity and unplanned surgical or interventional procedures in the interstage period. This study aimed to identify potential predictors of RAAO by analyzing initial echocardiograms and angiograms before hybrid stage 1 palliation. For this study, 96 patients who underwent hybrid stage 1 palliation between July 2002 and July 2009 were reviewed, 68 of which had standard HLHS and met the inclusion criteria. The initial echocardiogram, hybrid stage 1 angiograms, and follow-up echocardiograms were reviewed. Anatomic and hemodynamic measurements were obtained by both modalities, and comparisons were made between those who developed RAAO and those who did not. Of the 68 patients, 20 (29%) had RAAO. The mean aortic root size was smaller for the patients who had RAAO (3.6 vs 4.4 mm; p = 0.036). The angiographic angle between the aortic isthmus and the patent ductus arteriosus (PDA) was significantly larger in the RAAO group (86° vs 63°; p = 0.008). The retrograde aortic arch velocities were higher in the RAAO group. Patients with RAAO have a smaller aortic root and higher retrograde velocities on initial echocardiogram. Patients with RAAO show a larger angle between the retrograde arch and PDA on angiogram. Because RAAO is an important potential complication after hybrid stage 1 palliation for HLHS, identification of predictors of RAAO may lead to improved care and outcome for patients with RAAO.

Keywords

Hybrid stage 1 palliation; Hypoplastic left heart syndrome; Retrograde aortic arch obstruction

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The hybrid approach to palliation of hypoplastic left heart syndrome (HLHS) using pulmonary artery bands, stent placement in the patent ductus arteriosus (PDA), and atrial septostomy has evolved as an alternative treatment strategy for patients with HLHS [3, 4]. Obstruction of retrograde flow in the aortic arch is a recognized complication.

Previous studies have demonstrated that the incidence of retrograde aortic arch obstruction (RAAO) is 10% to 24% [2, 4, 7]. As a complication, RAAO increases morbidity and mortality immediately postoperatively as well as during the "interstage" period before the patient undergoes the second stage of palliative surgery or comprehensive stage 2 repair consisting of bidirectional cavopulmonary anastomosis, removal of the pulmonary artery

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bands and PDA stent, Damus-Kaye-Stansel procedure, aortic arch reconstruction, and atrial septectomy.

Retrograde aortic arch flow is responsible for perfusion of the head and neck vessels, and the coronary arteries. In HLHS patients with aortic atresia, this flow is particularly important because it is the sole supply of perfusion to these vital organs. Due to the importance of this flow, RAAO often leads to additional interstage procedures.

Various strategies have been used to treat obstruction after it is recognized. Examples include interventional catheterization procedures such as balloon angioplasty and stent placement in the retrograde aortic arch, reverse aorticopulmonary shunt, conversion to Norwood palliation, and advancement to comprehensive stage 2 repair [1, 7]. If the obstruction persists, decreased ventricular function and increased tricuspid regurgitation may develop.

Morphologic predictors associated with the development of RAAO after PDA stent placement as a part of hybrid stage 1 palliation have not been studied. This study aimed to identify anatomic predictors of RAAO by analyzing initial echocardiograms and angiograms before hybrid stage 1 palliation.

Methods

Study Population

The study included patients with classic HLHS (aortic stenosis/atresia with accompanying mitral stenosis/atresia) who had undergone hybrid stage 1 palliation at a single institution between July 2002 and July 2009. During this time, 96 patients underwent hybrid stage 1 palliation. Of the 96 patients, 20 were excluded from the analysis due to underlying anatomy comprising a variant of other complex single- or two-ventricle anatomies. Inclusion in the analysis required that patients had follow-up echocardiographic data at least 1 month after palliation or had undergone a procedure for retrograde obstruction.

Six patients were excluded from the study because of inadequate follow-up assessment. One patient was excluded due to an unusual aortic arch branching pattern that did not allow for adequate measurements. One patient was excluded because the hybrid stage 1 palliation was performed at 93 days of life after transfer from an outside institution, much later than the typical time of the procedure.

Consequently, a total of 68 patients (47 boys) with a mean age of 7.5 days and weighing 3 kg at the time of palliation were included in the analysis. The study was approved by the institutional review board. Patient information was obtained from both the electronic medical record and the patient's paper chart when necessary.

Data Collection

Echocardiographic Measurements—The initial echocardiogram before hybrid stage 1 was reviewed, and anatomic measurements were made at the maximum dimension in systole (Figs. 1, 2). The echocar-diographic views used were the parasternal long axis view (A,B), the high parasternal/suprasternal notch view (C–F), and subxiphoid sagittal imaging (G), defined as follows:

A: Aortic valve annulus between the hinge points of the right coronary and the noncoronary cusps

B: Aortic root between the aortic valve annulus and the sinotubular junction

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C: Ascending aorta proximal to the innominate artery

D: Proximal transverse aortic arch between the innominate artery and the left common carotid artery

E: Distal transverse aortic arch at the base of the left common carotid artery

F: Aortic isthmus at the base of the left subclavian artery

G: Descending aorta at the diaphragm

If the echocardiogram did not clearly demonstrate an area of the aortic arch or if there was an abnormal branching pattern of the head and neck vessels, then the measurement for that particular area of the aorta was not recorded. Measurements were obtained by one researcher and validated by the senior investigator. The investigators were blinded to the patient group. Measurements were indexed to body surface area.

The measurements also were entered into an echocar-diographic reporting system, and z-scores for the aortic isthmus, ascending aorta, aortic root, and distal transverse aortic arch were obtained. The z-scores were based on the Children's Hospital of Boston normative database [6].

Follow-up echocardiograms were obtained routinely the day after hybrid stage 1 palliation. The patients then had subsequent studies every 2 weeks during the interstage period or more frequently as clinically indicated. Other echocardiographic measurements were assessed at initial and follow-up studies including peak Doppler velocity in the retrograde aortic arch, peak velocities through the PDA stent and pulmonary artery bands, qualitative right ventricular function, and degree of tricuspid regurgitation. The peak Doppler velocities in the retrograde aortic arch were categorized into groups of 3 m/s or less, 3.1 to 3.5 m/s, and more than 3.5 m/s (Fig. 3).

Angiographic Measurements—All angiograms obtained during PDA stent placement were reviewed. The lateral angiogram was used for the following measurements of the aortic arch at maximum dimension (Fig. 4):

A: Ascending aorta

B: Transverse aortic arch at the base of the left common carotid

C: Orifice of the aortic isthmus

D: Proximal descending aorta

The angle between the aortic isthmus and the ductal arch was measured before stent deployment and on the post-deployment lateral angiogram. Transecting lines were drawn through the middle of the vessels: one through the orifice of the aortic isthmus and one from the middle of the ductus arteriosus to the proximal descending aorta (Fig. 5). The positioning of the PDA stent in relation to the aortic isthmus and descending aorta was defined as proximal to the orifice of the retrograde aortic isthmus, in the middle of the orifice of the isthmus, or distal to the orifice of the isthmus into the descending aorta.

Two investigators blinded to the patient group performed the measurements independently. All angiographic measurements were compared and demonstrated good interobserver reliability with an intraclass correlation of 0.855 [5]. The retrograde aortic arch gradient, defined as the systolic blood pressure difference between the transverse aortic arch and the

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descending aorta, was recorded from the cardiac catheterization reports at the time of the subsequent interventional catheterization in the RAAO group.

Outcome Variables—The patients were divided into two groups. The first group consisted of patients who had undergone an unplanned surgical or transcatheter intervention for RAAO (RAAO group). This group was based on a previous description of RAAO, with the inclusion of more recently added patients during the study period [7]. The decision to proceed with an interstage intervention was based on several factors including echocardiographic changes, the patient's clinical status, and electrocardiogram alterations. Important echo-cardiographic information included increased retrograde aortic arch gradient, decreased right ventricular function, and increased tricuspid regurgitation. Clinical changes such as decreased weight gain, increasing blood pressure gradients between the upper and lower extremities, and evidence of ST segment changes on electrocardiogram also contributed to the decision to intervene. Clinical management decisions were made after discussion at our combined cardiology/cardiothoracic surgery case management conference. The second group of patients included those who had not undergone an interventional procedure for RAAO (non-RAAO group).

The primary outcome measures were comparisons of the aortic dimensions and angle of the ductus arteriosus with the isthmus between the RAAO and non-RAAO groups. The secondary outcome measures included comparison of the stent type, stent location, retrograde arch velocities, and HLHS anatomic subtypes between the two groups.

Statistical Analysis—Continuous-variable measurements were compared between the patients with and those without RAAO using *t* tests. Paired *t* tests were used to compare measurements made with echocardiogram and angiogram. Differences in categorical measurements such as stent type and placement were made using chi-square and confirmed by Fisher's exact tests when cell sizes were small. To assess retrograde and PDA velocity over time, ordinary least squares was used to estimate unknown parameters in a linear regression model, with clustering of multiple measurements per patient over time to correct for standard errors.

Results

Of the 68 patients, 20 (29%) had RAAO. Complete echo-cardiographic data, including all necessary views before palliation, were available for 60 of the 68 patients, and complete angiographic studies were available for 57 of the 68 patients. No significant difference in age or weight at hybrid stage 1 or type of PDA stent between the RAAO and non-RAAO groups was observed (Tables 1, 2).

The two groups also were compared based on their underlying anatomy of aortic atresia/ mitral atresia, aortic atresia/mitral stenosis, and aortic stenosis/mitral stenosis. The mean ascending aorta measurement in the aortic stenosis/mitral stenosis group was 4.3 ± 1.1 mm (z-score, -2.1 ± 0.8). This was significantly larger than in the patients with aortic atresia, whose mean ascending aorta measurement was 2 ± 0.8 mm (z-score, –3.9 ± 0.7; $p < 0.001$). The patients with aortic stenosis/mitral stenosis typically had small jets of antegrade blood flow through the stenotic aortic valve. Of the 18 patients in this subgroup, 7 had antegrade aortic flow extending into the midportion of the ascending aorta, with the remaining 11 having retrograde flow. A trend toward a higher percentage of RAAO was found in the groups with aortic atresia (Table 3). The patients with aortic atresia underwent intervention for RAAO at a mean of 78 ± 56 days. The patients in the aortic stenosis/mitral stenosis group had intervention for RAAO at a mean of 96 ± 83 days.

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Echocardiography Results

The measurements of the aortic root were significantly smaller in the RAAO group. The aortic root measured 3.6 ±1.3 mm in the RAAO group, which was significantly smaller than in the non-RAAO group (4.4 ±1.4 mm) ($p = 0.036$). The z-scores of the aortic root also were significantly smaller in the RAAO group (–3.1 ± 0.8) than in the non-RAAO group (–2.5 ± 0.9) ($p = 0.024$).

Although the mean measurements and z-scores of the aortic annulus, ascending aorta, proximal transverse aortic arch, and aortic isthmus were smaller in the RAAO group, these differences did not reach statistical significance. The measurements of the aortic annulus and the aortic isthmus were significantly smaller in the RAAO group than in the non-RAAO group when indexed to body surface area (Table 4). A logistic regression analysis demonstrated that the aortic root z-score had an odds ratio of 0.43 (p = 0.03), suggesting that for every z-score increase of one standard deviation, the risk of RAAO decreases by 57%. The aortic arch measurements of the patients with aortic atresia also were compared between the RAAO and non-RAAO groups, and no significant differences were observed within this subgroup of patients (Table 5).

The follow-up echocardiograms demonstrated that retrograde aortic arch Doppler velocities increased over time in both groups of patients. The peak retrograde velocities were distributed differently between the groups, with higher velocities in the RAAO group ($p = 0.036$). For example, 20% of the patients in the RAAO group had velocities greater than 3.5 m/s compared with 2% of the non-RAAO group (Table 6). The RAAO group also had slightly higher (0.1 m/s) retrograde velocities than the non-RAAO group before hybrid palliation. Furthermore, the RAAO group increased velocity in the retrograde aortic arch more rapidly over time, with an additional 0.004 m/s per day more than the non-RAAO group ($p = 0.017$) (Fig. 6).

All four patients whose initial retrograde arch velocity was 2 m/s or more were in the RAAO group. The analysis demonstrated no significant differences during the interstage period between the two groups regarding right ventricular function, degree of tricuspid regurgitation, or velocity through the PDA stent (Fig. 7).

Angiographic Results—No significant differences in the angiographic measurements of the ascending aorta, transverse aortic arch, aortic isthmus, or descending aorta were observed between the RAAO and non-RAAO groups (Table 7a). In the subgroup of patients with aortic atresia, the ascending aorta and transverse aortic arch were smaller in the group that had RAAO (Table 7b). No significant differences between the RAAO and non-RAAO groups were observed with regard to PDA stent position related to the aortic isthmus (Table 8).

The angle on the initial angiogram between the aortic isthmus and the PDA was significantly greater in the RAAO group (86° ± 28°) than in the non-RAAO group (63° ± 29°) ($p = 0.008$). Stent deployment did not lead to a significant change in the angle (Table 9). Further analysis showed that the patients with an echocardiographic aortic root z-score of –3 or less and an angiographic angle of 60° or more experienced RAAO 53% of the time.

Comparison of Echocardiographic and Angiographic Measurements—The measurements of the ascending aorta, transverse aortic arch, aortic isthmus, and descending aorta were compared between the two methods. The aortic isthmus measured larger on the angiogram (4.3 ± 0.9 mm) than on the echocardiogram (3.9 ± 0.9 mm) ($p \leq 0.001$). The measurements of the ascending aorta, transverse aortic arch, and descending aorta were not statistically different (Table 10).

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The retrograde aortic arch gradient measured in the catheterization laboratory was not significantly different from the most recently estimated gradient on echocardiogram. The retrograde aortic arch pressures were obtained during cardiac catheterization using a 0.014-in. Radi (St. Jude Medical, St. Paul, MN) or Volcano (Volcano Corporation, San Diego, CA) wire, which has a pressure transducer near the distal tip. The pressure wire helps to eliminate a dampening effect from crossing a narrow lumen vessel with a catheter that has a larger diameter than the wire, resulting in an artificially lower pressure gradient. In the limited patient population that underwent an interventional cardiac catheterization for RAAO, the estimated echocardiographic gradient was 35 ± 15 mmHg compared with 31 ± 3 mmHg for catheterization with the patient under general anesthesia ($p = 0.15$).

Discussion

Retrograde aortic arch obstruction after hybrid stage 1 palliation for HLHS may lead to increased morbidity and often requires additional interstage interventional or surgical procedures [7]. This retrospective analysis evaluated the initial echocardiograms at presentation and the angiograms at the time of the hybrid stage 1 palliation in an attempt to identify predictors of RAAO. A slightly higher percentage of patients had a diagnosis of RAAO (29%) than in previous reports (10–24%). This may be secondary to selection bias in the study design using a specific subgroup of the overall population that underwent hybrid stage 1 palliation.

This study demonstrated that patients who had RAAO started out with a smaller aortic root size than the non-RAAO group. Patients who comprised the aortic stenosis/mitral stenosis subgroup of HLHS had RAAO less frequently than the patients with aortic atresia. Even if these patients did experience increased velocity in the retrograde aortic arch, the antegrade flow supplying the coronary arteries could temper the effect of RAAO and prevent the need for an additional interstage procedure.

Although the data from the follow-up echocardiograms demonstrated that the velocities through the PDA stent and retrograde aortic arch routinely increased in all patients over time, the RAAO patients started with a slightly higher velocity in the retrograde aortic arch that increased at a slightly faster rate over time than in the non-RAAO group. Clinically, these data support the idea that not every patient with increasing velocity in the retrograde aortic arch requires intervention. However, patients with more rapid increases in velocity or evidence of other secondary effects, such as worsening right ventricular function or tricuspid regurgitation, should be considered for intervention. Also, all four of the patients who had a velocity in the retrograde aortic arch of 2 m/s or more on the initial echocardiogram received an intervention for RAAO.

The angle between the ductus arteriosus and the aortic isthmus was higher in the group that had RAAO, which could alter the dynamics of the blood flow into the retrograde aorta and the vital organs that it perfuses. Stent implantation did not lead to an immediate change in the angle. This study, however, did not evaluate for changes in this angle on repeat cardiac catheterization that could occur as the patients grow.

In addition to the patients with a retrograde velocity of 2 m/s or greater, another patient population was identified as having a higher risk for the development of RAAO. The patients with an aortic root z-score of –3 or less and an angiographic angle of 60° or more experienced RAAO 53% of the time. This was nearly twice the likelihood of the total group risk.

Overall, the measurements obtained by echocardiography correlated well with the angiographic results. Although a significant difference in the measured size of the aortic

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isthmus was found between the methods, the absolute difference was small. This area is a difficult area to image by both echocardiography and angiography. Increased experience with hybrid stage 1 palliation led to better recognition of the importance of demonstrating this region of the aorta clearly before palliation. Despite the differences in measurements between modalities, there was no significant difference in aortic isthmus measurements when comparing between the RAAO and non-RAAO groups by either echocardiography or angiography.

One limitation of the study is that it was a retrospective analysis with incomplete images available for analysis, especially from cases earlier in the experience of our center. Although this is the largest series reported, another limitation is the relatively small study group size, which limits the ability to perform certain subgroup analyses. Also, because the data were analyzed retrospectively, right ventricular function was qualitatively assessed as opposed to the use of more quantitative methods.

The data demonstrate that the RAAO group had a higher percentage of patients with velocities greater than 3.5 m/s. As previously noted, the decision to perform an intervention is not based solely on the retrograde velocity. Many other echocardiographic findings, electrocardiogram changes, and clinical findings contribute to the decision whether to intervene for RAAO. The retrograde velocity may not truly reflect some cases of obstruction if concomitant worsening of the right ventricular function occurs. In our study, one patient who had evidence of severe RAAO on echocardiogram did not have evidence of RAAO at catheterization, and no intervention was required.

Further evaluation of these patients is required to determine the ultimate basis for the development of RAAO after hybrid stage 1 palliation. This study suggests that the size of the aortic root and the angle the aortic isthmus arising from the PDA may be anatomic factors contributing to RAAO. The initial velocity in the retrograde aortic arch on echocardiogram is another factor associated with the development of RAAO. Further analysis of the changes in the angle between the aortic isthmus and the PDA over time with somatic growth as well as attempts to measure this angle on echocardiogram could be areas of future research.

A small percentage of patients with HLHS are born with a congenital anatomic variant of severe RAAO that manifests prenatally or shortly after delivery. When this rare form of the anatomy is recognized, it is a contraindication to hybrid stage 1 palliation [7]. The anatomic factors found to be significantly associated with the development of RAAO in this study have not altered the selection criteria for hybrid stage 1 palliation for patients with HLHS at our institution. However, the results identified a high-risk subgroup of patients that demand vigilant follow-up assessment to evaluate for clinical and echocardiographic changes of RAAO. As with all patients, when indications of RAAO arise, we maintain a low threshold for hemo-dynamic and interventional cardiac catheterization.

Additionally, studies to evaluate the histopathologic response of the surrounding tissue to the PDA stent are ongoing. There may be a subset of patients who have a heightened immunologic response to stent placement, which could lead to obstruction of flow in the retrograde aortic arch as well. Because the etiology of RAAO likely is multifactorial, it continues to require a high index of suspicion to prevent its possible sequelae.

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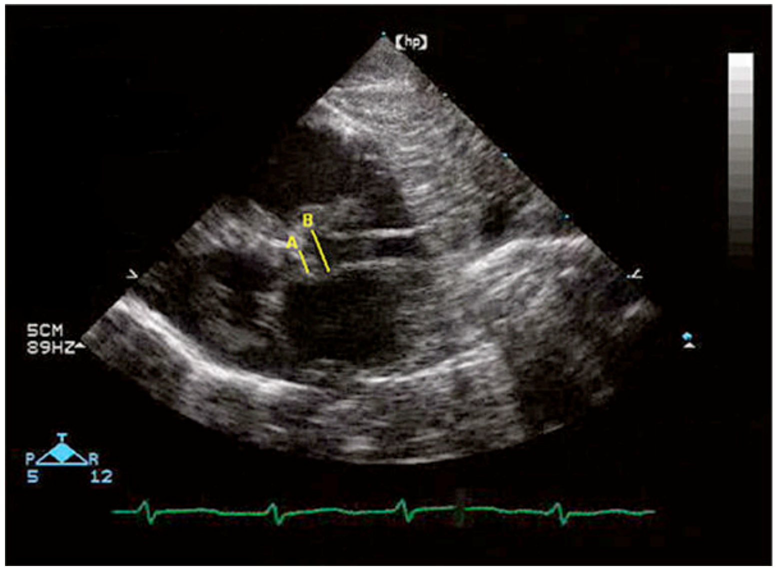

Fig. 1.

Aortic annulus and aortic root measurements. This parasternal long-axis view shows measurements of the aortic annulus (*A*) and aortic root (*B*)

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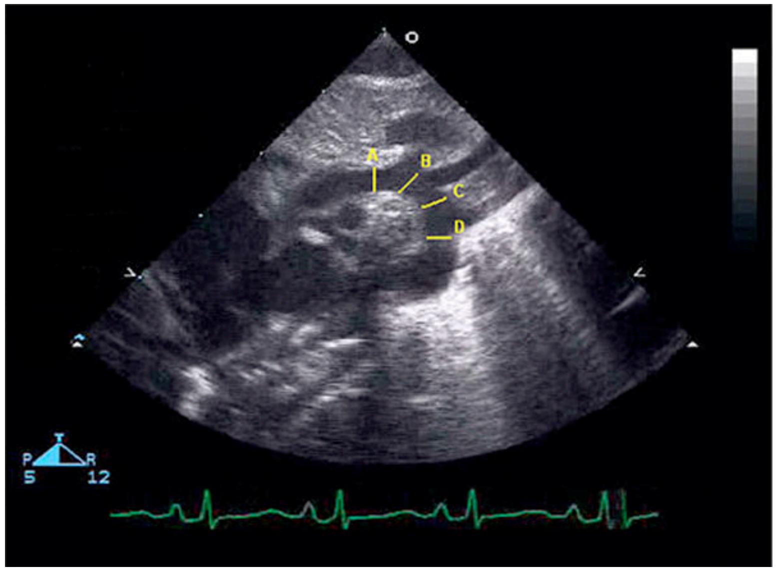

Fig. 2.
Aortic arch measurements. This high parasternal view shows measurements of the ascending aorta (*A*), proximal transverse aortic arch (*B*), distal transverse aortic arch (*C*), and aortic isthmus (*D*)

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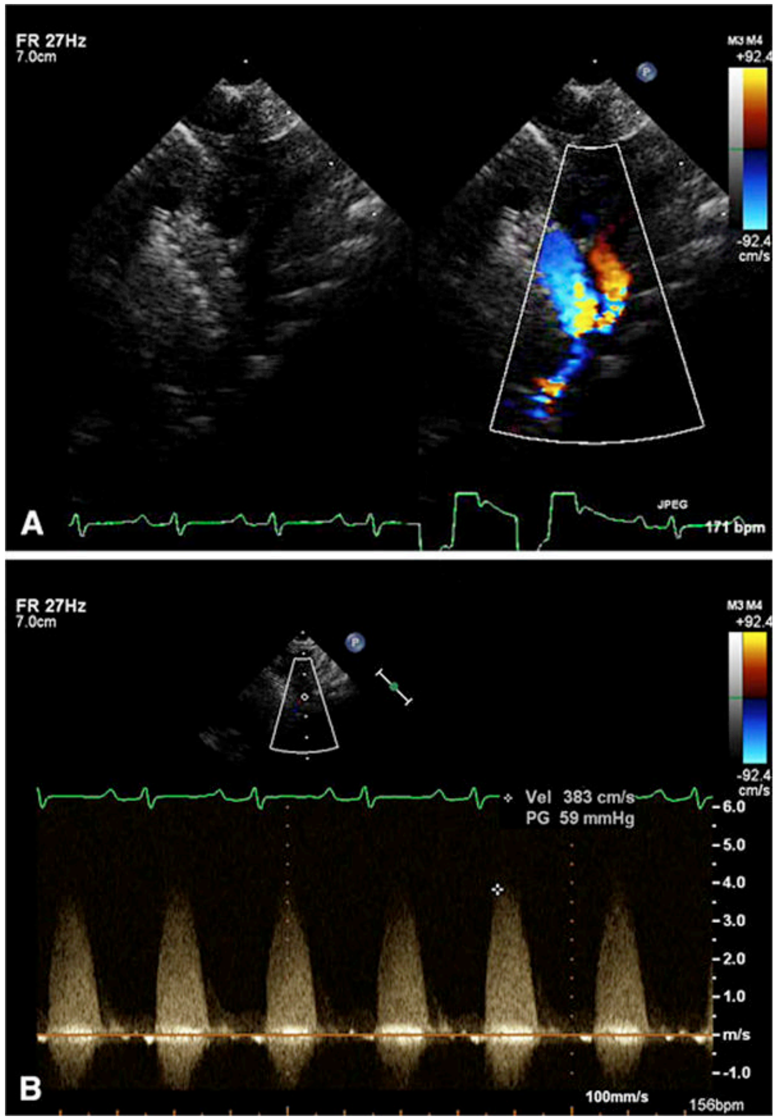

Fig. 3.

a Patent ductus arteriosus (PDA) stent seen by two-dimensional imaging (*left*) and color Doppler (*right*) demonstrating antegrade flow through the PDA stent. There is aliased retrograde flow from the PDA stent into the retrograde aortic arch. **b** Continuous-wave Doppler analysis of retrograde aortic arch flow in a patient with retrograde aortic arch obstruction (RAAO)

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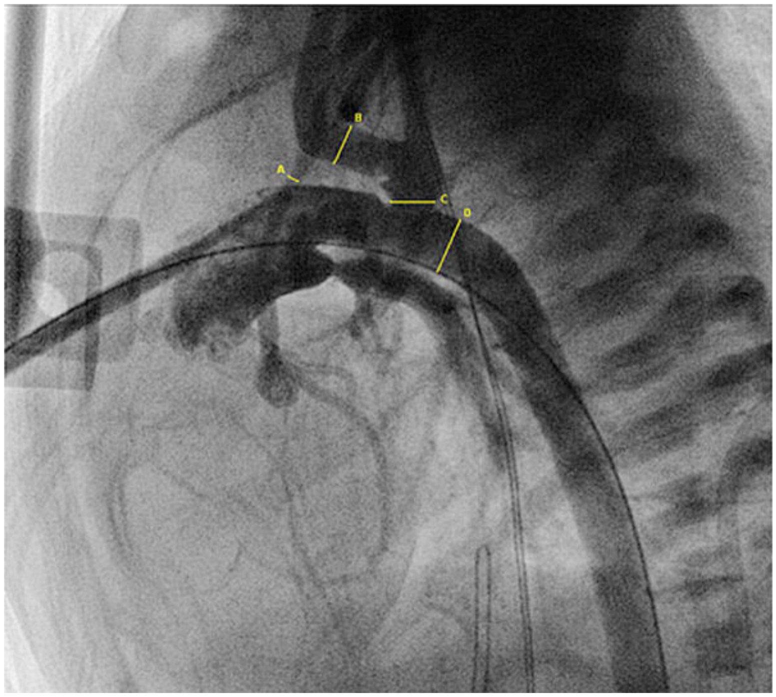

Fig. 4.
Lateral angiogram obtained by an injection through a 6-Fr sheath placed in the main pulmonary artery during hybrid stage 1 palliation demonstrating the measurements of the ascending aorta (*a*), transverse aortic arch (*b*), aortic isthmus (*c*), and descending aorta (*d*)

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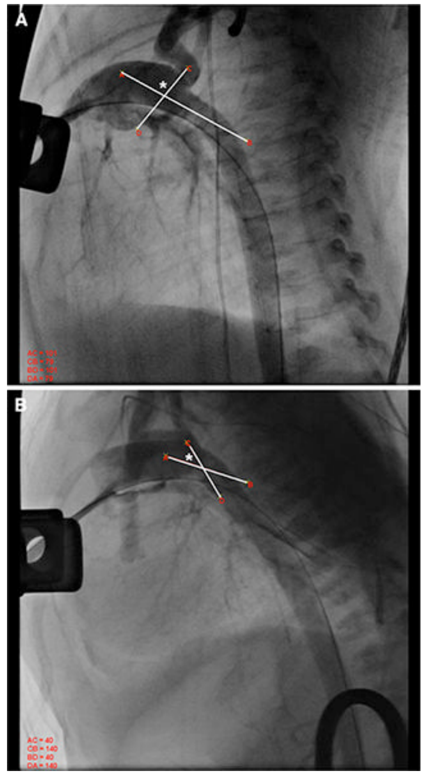

Fig. 5.

Two measurements of the angle between the aortic isthmus orifice and the ductus arteriosus are demonstrated. The angle is obtained by intersecting lines that transect the patent ductus arteriosus (PDA) and the entrance of the aortic isthmus (*). **a** Angle of 101°. **b** Angle of 40° between the aortic isthmus and the patent ductus arteriosus (PDA)

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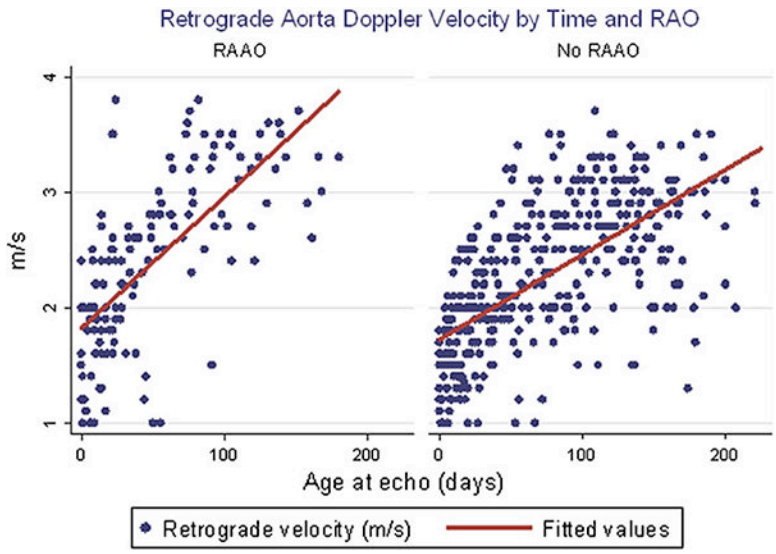

Fig. 6.

The retrograde aortic arch obstruction (RAAO) group started with slightly higher retrograde aortic arch velocities that increased at a slightly faster rate over time than the group that did not experience RAAO, as demonstrated by the slope of the line

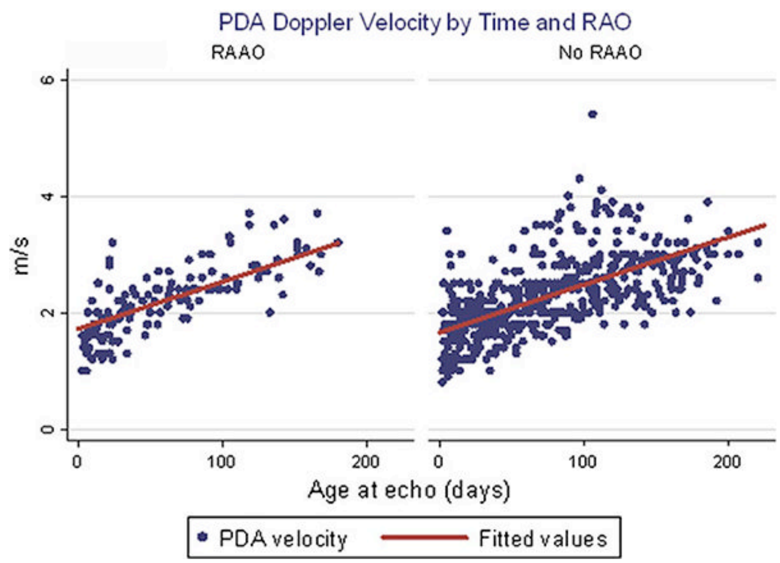

Fig. 7.
The patent ductus arteriosus (PDA) velocities increased over time in both groups at similar rates

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Table 1

Demographics

	RAAO	Non-RAAO	*p* Value
Age at hybrid (days)	8 ± 5.6	7.3 ± 4.2	0.581
Weight (kg)	3.1 ± 0.8	2.9 ± 0.6	0.297
Male/female	13/7	34/14	0.635

RAAO retrograde aortic arch obstruction

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Table 2

Stent type

Stent type	RAAO (%)	Non-RAAO (%)	*p* Value
Balloon expandable	33	67	0.771
Self-expandable	29	71	

RAAO retrograde aortic arch obstruction

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Table 3

Anatomy

Anatomy	RAAO (%)	Non-RAAO (%)	*p* Value
AS/MS (*n* = 18)	17	83	0.326
AA/MA (*n* = 32)	31	69	
AA/MS (*n* = 18)	39	61	

RAAO retrograde aortic arch obstruction, *AS* aortic stenosis, *MS* mitral stenosis, *AA* aortic atresia, *MA* mitral atresia

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Table 4

Echocardiographic measurements of the aorta in all patients

		RAAO (mm)	Non- RAAO (mm)	*p* Value
Aortic annulus		2.3 ± 1	2.8 ± 0.8	0.066
	z-score	–3.9 ± 0.8	–3.5 ± 0.8	0.065
	Index BSA	5 ± 2.1	6.3 ± 1.9	0.041
Aortic root		3.6 ± 1.3	4.4 ± 1.4	0.036
	z-score	–3.1 ± 0.8	–2.5 ± 0.9	0.024
	Index BSA	7.7 ± 2.7	9.9 ± 3.3	0.018
Ascending aorta		2.2 ± 1.2	2.7 ± 1.4	0.170
	z-score	–3.7 ± 0.8	–3.3 ± 1.1	0.128
	Index BSA	4.8 ± 2.3	6.2 ± 3.3	0.105
Proximal transverse		3.7 ± 0.6	3.9 ± 0.9	0.401
aortic arch	Index BSA	8.1 ± 1.2	8.6 ± 1.8	0.260
Distal transverse		4.2 ± 0.8	4.1 ± 0.9	0.699
aortic arch	z-score	–2.2 ± 0.5	–2.3 ± 0.5	0.943
	Index BSA	9.1 ± 1.8	9.1 ± 1.8	0.957
Aortic isthmus		3.6 ± 0.6	4 ± 1	0.108
	z-score	–2.5 ± 0.4	–2.1 ± 0.8	0.059
	Index BSA	7.7 ± 1.1	8.9 ± 2.2	0.045
Descending aorta		6.3 ± 1.2	5.9 ± 0.7	0.127
	Index BSA	13.6 ± 2.1	13 ± 1.6	0.237

RAAO retrograde aortic arch obstruction, *BSA* body surface area

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Table 5

Echocardiographic measurements of the aorta in aortic atresia

		RAAO (mm)	Non-RAAO (mm)	*p* Value
Aortic annulus		2 ± 0.8	2.5 ± 0.7	0.167
	z-score	–4 ± 0.7	–3.8 ± 0.8	0.346
	Index BSA	4.6 ± 1.6	5.4 ± 1.5	0.170
Aortic root		3.3 ± 0.9	3.8 ± 1.2	0.164
	z-score	–3.2 ± 0.6	–2.9 ± 0.8	0.230
	Index BSA	7.1 ± 1.9	8.4 ± 2.8	0.144
Ascending aorta		1.8 ± 0.4	2 ± 0.9	0.427
	z-score	–3.9 ± 0.4	–3.8 ± 1.1	0.619
	Index BSA	4 ± 0.8	4.5 ± 2.1	0.404
Proximal transverse		3.7 ± 0.6	3.8 ± 0.8	0.772
aortic arch	Index BSA	8.2 ± 1.2	8.3 ± 1.7	0.884
Distal transverse		4 ± 0.8	4.1 ± 0.9	0.803
aortic arch	z-score	–2.3 ± 0.5	–2.3 ± 0.6	0.801
	Index BSA	8.8 ± 1.7	8.9 ± 1.8	0.864
Aortic isthmus		3.6 ± 0.6	3.8 ± 0.8	0.416
	z-score	–2.5 ± 0.5	–2.3 ± 0.7	0.390
	Index BSA	7.7 ± 1.2	8.2 ± 1.8	0.360
Descending aorta		6.2 ± 1.2	5.8 ± 0.7	0.224
	Index BSA	13.5 ± 2.1	12.8 ± 1.6	0.209

RAAO retrograde aortic arch obstruction, *BSA* body surface area

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Table 6

Peak retrograde aortic arch velocities

Retrograde velocity	RAAO (n = 20) n (%)	Non-RAAO (n = 47) n (%)	p Value
≤3 m/s	11 (55)	29 (62)	0.036
3.1–3.5 m/s	5 (25)	17 (36)	
>3.5 m/s	4 (20)	1 (2)	

RAAO retrograde aortic arch obstruction

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Table 7

Angiographic measurements

	RAAO (mm)	Non-RAAO (mm)	*p* Value
(a) In all patients			
Ascending aorta	2.3 ± 1.4	2.6 ± 1.1	0.375
Transverse aortic arch	3.9 ± 0.6	4.3 ± 0.8	0.107
Aortic isthmus	4.3 ± 1.1	4.4 ± 0.8	0.684
Descending aorta	6.1 ± 0.8	6.2 ± 0.9	0.615
(b) In patients with aortic atresia			
Ascending aorta	1.8 ± 0.4	2.3 ± 0.9	0.038
Transverse aortic arch	3.8 ± 0.6	4.4 ± 0.8	0.015
Aortic isthmus	4.1 ± 1	4.4 ± 0.9	0.308
Descending aorta	5.8 ± 0.7	6.2 ± 0.9	0.105

RAAO retrograde aortic arch obstruction

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Table 8
Placement of the patent ductus arteriosus (PDA) stent in relation to the aortic isthmus

Placement	RAAO (%)	Non-RAAO (%)	*p* Value
Proximal ($n = 6$)	17	83	0.098
Mid ($n = 19$)	16	84	
Distal ($n = 33$)	42	58	

RAAO retrograde aortic arch obstruction

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Table 9

Aortic isthmus–ductus arteriosus angle measurements

Isthmus angle	RAAO (°)	Non-RAAO (°)	*p* Value
Pre-stent	86 ± 28	63 ± 29	0.008
Post-stent	80 ± 25	66 ± 28	0.064

RAAO retrograde aortic arch obstruction

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Table 10

Comparison of measurements by method

	Echocardiogram (mm)	Angiogram (mm)	*p* Value
Ascending aorta	2.6 ± 1.4	2.5 ± 1.2	1.000
Transverse aortic arch	4.1 ± 0.8	4.2 ± 0.8	0.136
Aortic isthmus	3.9 ± 0.9	4.3 ± 0.9	<0.001
Descending aorta	6.0 ± 0.9	6.1 ± 0.9	0.446