Surgical Management of Severe Aortic Outflow Obstruction in Lesions Other Than the Hypoplastic Left Heart Syndrome: Use of a Pulmonary Artery to Aorta Anastomosis

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Between December 1985 and April 1990, 50 infants with a variety of congenital cardiac lesions other than the hypoplastic left heart syndrome underwent surgical relief of aortic outflow obstruction by creation of a pulmonary artery to aorta anastomosis. The patients were grouped anatomically by ventriculoarterial alignment. Nineteen had normally aligned great arteries (group I); 25 had transposition of the great arteries, all with a bicuspid left ventricular heart of left ventricular morphology (group II); and 6 had a double-outlet right ventricle (group III). All patients had either aortic stenosis with atresia, subaortic stenosis or a restrictive ventricular septal defect. Sixteen had normal arch anatomy; 34 had arch anomalies consisting of arch hypoplasia (n = 17), coarctation (n = 11), interruption of the arch (n = 4) and complex arch anomalies (n = 2).

Surgery was performed at a median age of 10 days (range 2 to 184). Of the 50 infants, 33 survived. No significant difference in early survival (30 days) was noted among the groups of varying ventriculoarterial alignment (68% group I, 72% group II, 83% group III; p = 0.05). Overall actuarial survival was 63% at 18 months. Analysis of actuarial survival by arch anatomy, although not statistically significant, revealed a trend toward better survival at 18 months postoperatively in infants with normal arch anatomy (81%) than in infants with arch anomalies (54%). Of the 33 survivors, 26 have proceeded to the next surgical stage, including the Fontan procedure in 8, superior cavopulmonary anastomosis in 13 and biventricular repair in 5.

Equal early survival among patients in various anatomic groups of ventriculoarterial alignment can be successfully achieved after performance of a pulmonary artery to aorta anastomosis in infancy for severe aortic outflow obstruction. Arch anatomy appears to correlate with survival, with a higher mortality rate seen in infants with aortic arch abnormalities.

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Methods

Study patients. We reviewed the records of all 50 infants who 1) did not have a diagnosis of hypoplastic left heart syndrome, and 2) underwent a pulmonary artery to aorta anastomosis procedure for relief of aortic outflow obstruction at the Children's Hospital of Philadelphia between December 1985 and April 1990. Surgery was performed at a median age of 10 days (range 2 to 184). Thirty-four of the infants were male and 16 were female.

Anatomic groups. The patients were classified into three anatomic groups according to ventriculoarterial alignment. These groups were as follows.

Group I. Nineteen patients had normally aligned great arteries, segments (S,D,S), with both two ventricles present. Aortic outflow obstruction consisted of subaortic stenosis, aortic stenosis or aortic atresia. In the past we have defined "hypoplastic left heart syndrome" as a mitral valve anulus <7 mm in diameter (measured by echocardiography in the four-chamber apical or subcostal short-axis view) and a short-axis diastolic left ventricular dimension <10 mm in diameter. Therefore, patients with mitral hypoplasia or atresia, aortic outflow obstruction, intact ventricular septum and hypoplasia of the left ventricle have been previously reviewed (9) and were excluded from this study. In the present review, all 18 patients in group I had a large, unrestrictive, ventricular septal communication and thus had a left ventricle that was not hypoplastic. Of the 19 patients, 8 had a normal size mitral valve and were assessed as having a left ventricle extending at least two thirds of the distance to the apex of the heart. These patients were thought to be suitable candidates for "biventricular" repair.

Group II. Twenty-five infants had transposition of the great arteries and univentricular heart of left ventricular morphology. Sixteen patients had an "L-loop" ventricular arrangement (segments S.L.L), double-inlet left ventricle with l-transposition of the great arteries. 4 of whom had left-sided atrioventricular (AV) valve atresia. Nine patients had a "D-loop" ventricular arrangement (segments D.S.D), all of whom had right AV valve atresia (tricuspid atresia with d-transposition of the great arteries). All 25 infants had a restrictive ventricular septal defect (see defining criteria).

Group III. Six infants had double-outlet right ventricle. All six had two ventricles; five of the six had a D-loop and 1 had an L-loop ventricular arrangement. All had an unrestrictive ventricular septal defect, with subaortic or aortic stenosis, or both, as the cause of aortic outflow obstruction.

A wide spectrum of aortic arch anatomy was seen. In total, 16 patients had normal arch anatomy and 34 had either aortic arch hypoplasia, coarctation of the aorta or interruption of the aortic arch, type B (Table 1). Two of the 50 infants had double aortic arch consisting of a right cervical aortic arch and atresia of the left arch.

Preoperative evaluation and interventions. All 50 infants underwent detailed two-dimensional echocardiographic evaluation of anatomy before operation. In the presence of a nonrestrictive ductus arteriosus, the presence or absence of a pressure gradient is not a reliable indicator of the severity of aortic obstruction. Thus, aortic anulus diameter or ventricular septal defect size was measured. Aortic anulus size <6 mm in diameter or ventricular septal defect orifice size smaller than that of the aortic anulus was considered restrictive.

Although cardiac catheterization was not routinely performed, 16 patients (8 at the referring institution) underwent the procedure before creation of a pulmonary artery to aorta anastomosis. Reasons for preoperative cardiac catheterization included the surgeon's wish to obtain additional images of aortic arch anatomy or angiographic delineation of left ventricular size.

One patient had undergone pulmonary artery banding at another institution at age 1 month before referral to our institution for a pulmonary artery to aorta anastomosis. Reasons for preoperative cardiac catheterization included the surgeon's wish to obtain additional images of aortic arch anatomy or angiographic delineation of left ventricular size.

Peripheral perfusion was restored in all patients.

Surgical technique. In infants with a small mitral valve, an unrestricted interatrial communication was created by excision of the septum primum. The pulmonary artery was divided at the origin of the right and left branches and the distal main pulmonary artery was oversewn with a patch. In infants with coarctation of the aorta or aortic arch hypoplasia, the approach was similar to that described by Norwood et al. (11) for palliation of hypoplastic left heart syndrome. A wide aortotomy was made from the thoracic aorta to the level of the transected proximal main pulmonary artery. A gusset of patch material made from pulmonary homograft was used to reinforce the aortotomy site. The homograft gusset was then sutured to the ascending aorta at the referring institution.

Table 1. Distribution of Arch Anatomy Among the 50 Infants

<table>
<thead>
<tr>
<th>Arch Anatomy</th>
<th>No. (%)</th>
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<tbody>
<tr>
<td>Normal</td>
<td>16 (32)</td>
</tr>
<tr>
<td>Arch hypoplasia</td>
<td>17 (34)</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>11 (22)</td>
</tr>
<tr>
<td>Interruption of the aortic arch type B*1</td>
<td>4 (8)</td>
</tr>
<tr>
<td>Complex arch anomaly</td>
<td>2 (4)</td>
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*Celoria and Patton classification.
Figure 1. Pathologic specimen from a patient after surgical palliation of transposition of the great arteries (S.L.L.), double-inlet left ventricle, mildly hypoplastic left atrioventricular valve and aortic outflow obstruction due to a restrictive ventricular septal defect. The white arrowhead indicates pulmonary outflow, the black arrowhead indicates aortic outflow. 1 = the wide open interatrial communication after excision of the septum primum; 2 = the left ventricle; 3 = the small right ventricular outflow chamber made that the restrictive ventricular septal defect is smaller than the aortic valve annulus; 4 = the anastomosis between the pulmonary arteries and the aorta with pulmonary homograft augmentation; 5 = the central shunt from the reconstructed aortic arch to the pulmonary arteries; 6 = the pulmonary arteries.

A 4-mm polytetrafluoroethylene tube graft was interposed between the underside of the aortic arch (central shunt or base of the right innominate artery [modified right Blalock-Taussig shunt]) and the branch pulmonary arteries to supply pulmonary blood flow. Of our patients, 38 had placement of a central shunt (81%) and 9 had a modified right Blalock-Taussig shunt (19%). The other three patients, who underwent primary biventricular repair, did not receive a shunt.

Eight infants in group 1 with aortic outflow obstruction and a large ventricular septal defect had a normal size mitral valve and left ventricle. Of these, three underwent primary ("biventricular") repair consisting of a pulmonary artery to aorta anastomosis in conjunction with patch closure of the ventricular septal defect to the native pulmonary mean (normal) valve, right ventriculotomy and placement of a 10-mm conduit from the right ventricle to the branch pulmonary arteries. The other five underwent initial pulmonary artery to aorta anastomosis with biventricular repair performed at a subsequent date.

Statistical methods. Numeric values were expressed as mean values ± SEM unless otherwise stated. Fisher's exact test was used in comparing early survival among the variables analyzed. Conventional life table analysis was used to construct actuarial survival curves for the various groups. Eighteen months was chosen as the cutoff period for analysis of survival after the initial palliative procedure because almost all survivors will have proceeded to the next surgical stage by then. The Tarone-Ware test was used to determine significance between the actuarial survival curves. A p value <0.05 was considered significant.

Results
Survival. Thirty-three of the 50 infants who underwent a pulmonary artery to aorta anastomosis survived. Postoperative follow-up until the next surgical stage or until the end of the study period (April 1990) ranged from 1 to 31 months. Actuarial survival of all 50 infants is shown in Figure 2; 63 patients survived to 18 months postoperatively.

Early survival was defined as up to 30 days after the initial palliative procedure. Seventy-two percent of infants survived the early postoperative period. Of the 17 deaths, 14 (82%) of all deaths were early (<30 days); 8 of these (47% of all deaths) were during the 1st 24 hours after operation. Causes of early death included acute cardiopulmonary collapse (n = 12), reconstructed aortic arch dissection at the site of patch homograft augmentation (n = 11) and intraoperative death with inability to wean from cardiopulmonary bypass secondary to coronary air embolus (n = 1). Causes of the three late deaths (>30 days) after the initial palliative procedure included sepsis (n = 1) in a patient still hospitalized after surgery, sudden cardiac arrest after cardiac catheterization before the next surgical stage (n = 1) and sudden cardiac arrest at home (n = 1).
The following variables were analyzed:

1. Ventriculoarterial alignment. Sixty-eight percent (13 of 19) of infants with normally related great arteries (group I). 72% (18 of 25) of infants with transposition of the great arteries (group II) and 83% (5 of 6) of infants with double-outlet right ventricle (group III) survived to 30 days after a pulmonary artery to aorta anastomosis, with no significant difference in early survival noted among the three groups (p > 0.05). Analysis of actuarial survival curves to 18 months after the initial palliative procedure for the three different groups of ventriculoarterial alignment showed no significant difference in survival to the next surgical stage (p > 0.05).

2. Right versus left ventricle as dominant systemic pumping chamber. The proximal pulmonary artery functions as the systemic great vessel after creation of a pulmonary artery to aorta anastomosis. In 50% of our patients, the pulmonary artery arose from a morphologic left ventricle (group II, n = 25) and in 50% from a morphologic right ventricle (groups I and III, n = 25). No difference in early survival was found between patients utilizing a morphologic left ventricle (18 [72%] of 25) versus a morphologic right ventricle (18 [72%] of 25) as the dominant systemic pumping chamber.

3. Shunt type. No difference in early survival was noted between patients who received a central shunt (7 [78%] of 9) and those who received a modified right Blalock-Taussig shunt (28 [74%] of 38) (p > 0.05). Three patients underwent a biventricular repair in conjunction with a pulmonary artery to aorta anastomosis and did not receive a systemic to pulmonary shunt.

4. Arch anatomy. Although not statistically significant, a difference in early survival was appreciated between infants with normal arch anatomy and infants with abnormal arch anatomy. Thirteen (81%) of 16 infants with normal arch anatomy and 23 (68%) of 34 infants with structural arch abnormalities survived to 30 days (p = 0.26). Actuarial survival curves to 18 months after the initial palliative procedure were constructed for the 16 infants with normal arch anatomy and the 34 with arch abnormalities (Fig. 3). Divergence between the two curves is seen (p = 0.17), with apparently better survival noted in infants lacking arch abnormalities. Eighty-one percent of infants with normal arch anatomy and 54% of infants with arch abnormalities survived to 18 months. No additional mortality was incurred by infants with normal arch anatomy after the initial 30 days after operation.

Additional procedures performed between pulmonary artery to aorta anastomosis and the next surgical stage. Three patients underwent placement of an additional aorta to pulmonary artery shunt to increase pulmonary blood flow after admission for increasing cyanosis and cardiac catheterization evaluation of significant shunt stenosis. One patient had a 40 mm Hg gradient across the aortic arch from proximal to distal reconstruction and underwent intraoperative balloon dilation performed with use of a previously described technique (12) that successfully eliminated the gradient.

Hemodynamic and angiographic findings after pulmonary artery to aorta anastomosis. Twenty-six of the 33 survivors have undergone cardiac catheterization for evaluation of hemodynamic status and anatomy before their second stage surgical procedure (Table 2). Cardiac catheterization was performed at a median age of 13 months (range 7 to 33). No patient had a gradient from the systemic ventricle to the ascending neoaoorta reconstruction (Fig. 4 and 5). (One patient had a significant distal arch gradient, as previously mentioned.) Of the 26 patients, 2 had mild neoaoorta (native pulmonary valve) regurgitation and none had significant native aortic valve regurgitation detected on angiography.

| Table 2. Cardiac Catheterization Data of 26 Patients After Performance of a Pulmonary Artery to Aorta Anastomosis Before the Next Surgical Stage |
|--------------------|------------------|-----------------|
| **Hemodynamic Variables** | **Mean Values (SD)** |
| Hgb (g/100 ml) | 16.5 (2) |
| RA pressure (mm Hg) | 5 (2) |
| LA pressure (mm Hg) | 6 (2) |
| SV pressure (mm Hg) | 110 (16) (9) |
| RPA pressure (mm Hg) | 13 (3) |
| LPA pressure (mm Hg) | 14 (4) |
| RPPW pressure (mm Hg) | 14 (3) |
| LPVW pressure (mm Hg) | 14 (3) |
| Asc neo-Ao pressure (mm Hg) | 108 (14) (50) |
| Desc Ao pressure (mm Hg) | 107 (12) (50) |
| SVR (Wood U) | 17 (7) |
| PVR (Wood U) | 2.0 (1.2) |
| Aortic oxygen saturation (%) | 76 (6) |
| Qp/Qs | 1.0 (0.5) |

Asc neo-Ao = ascending neo-aortic; Desc Ao = descending aorta; Hgb = hemoglobin; LA = left atrium; LPA = left pulmonary artery; LPVW = left pulmonary venous wedge; PVR = pulmonary vascular resistance; Qp/Qs = ratio of pulmonary to systemic flow; RA = right atrium; RPA = right pulmonary artery; RPPW = right pulmonary venous wedge; SVR = systemic vascular resistance.
Figure 4. Anteroposterior view of a left ventricular angiogram from an infant with single left ventricle (LV), transposition of the great arteries and a restrictive ventricular septal defect after surgical palliation. LPA = left pulmonary artery; RPA = right pulmonary artery; RV = right ventricle.

(Fig. 6). One patient had moderate tricuspid valve regurgitation. Nine patients were noted to have alterations in pulmonary artery architecture, eight with branch pulmonary artery deformity and one with pulmonary hypoplasia. Pulmonary artery narrowing or size in itself was not considered a contraindication to proceeding with the next surgical stage because generous pulmonary artery augmentation from hilum to hilum is always performed at the next stage of planned surgery (13). No patient was denied the next stage of surgical intervention on the basis of hemodynamic or angiographic findings at the time of cardiac catheterization.

Subsequent surgery. To date, 26 patients have undergone an additional elective palliative or definitive surgical procedure. Five patients with normal ventriculoarterial alignment, normal size left ventricle and normal size mitral valve have had a "biventricular" repair consisting of patch closure of the ventricular septal defect to the neo-aortic (native pulmonary) valve, ligation of the systemic to pulmonary shunt and right ventriculotomy with placement of a conduit from the right ventricle to the pulmonary arteries. This was performed at a median age of 18 months (range 11 to 20). All five survived and are doing well at home.

As of May 1989, candidates for a modified Fontan operation first undergo an intermediate procedure consisting of ligation of the systemic to pulmonary shunt, generous augmentation of the branch pulmonary arteries and creation of a cavopulmonary anastomosis between the superior vena cava and augmented branch pulmonary arteries. This procedure is performed to remove ventricular volume overload at an early stage. Thirteen patients have undergone this "hemi-Fontan" procedure, which was performed at a median age of 12 months (range 4 to 21) with no deaths. Four of these

Figure 5. Left ventricular angiogram (lateral view) demonstrating a severely restrictive ventricular septal defect and the site of the pulmonary arteries to aorta anastomosis, which allows for unimpeded flow from the left ventricle (LV) to the aorta (Ao). PA = pulmonary artery; other abbreviation as in Figure 4.

Figure 6. Lateral view of angiogram of the reconstructed arch after performance of a pulmonary artery (PA) to aorta (Ao) anastomosis with pulmonary homograft augmentation of hypoplastic aortic arch.
patients have since undergone completion of the modified Fontan procedure by patch baffling of the inferior vena cava blood flow to the pulmonary arteries, and none of these four have died.

Eight patients treated before May 1989 directly underwent a modified Fontan procedure at a median age of 20 months (range 14 to 30). Three of the eight died; the other five are at home and doing well.

Discussion

Obstruction to aortic outflow may occur in the setting of complex congenital heart disease and may appear in infants with lesions other than the hypoplastic left heart syndrome. In one review (1), 7% of pathologic specimens from patients <1 year old included some form of subaortic stenosis or atresia, with death occurring predominantly within the 1st month of life. Experience over the years with the more common form of aortic outflow obstruction, hypoplastic left heart syndrome (9), has enabled us to offer this smaller subset of anatomically heterogeneous patients a staged surgical approach that employs pulmonary artery to aorta anastomosis as initial palliation, ultimately leading to the Fontan procedure.

Experience with pulmonary artery banding and pulmonary artery to aorta anastomosis. Surgical recruitment of the pulmonary artery to supply systemic flow was first suggested in 1975 by Damus (14), Kaye (15) and Stansel (16) for physiologic correction of transposition of the great arteries and later utilized by Yacoub and Radley-Smith (17) in 1976 as a means of bypassing subaortic obstruction in a patient with "single ventricle" and subaortic stenosis.

Recently some investigators (18-22) have advocated a pulmonary artery to aorta anastomosis instead of resection of muscle or aortic arch conduit placement in patients who have developed subaortic stenosis after placement of a pulmonary artery band. In univentricular lesions, aortic outflow obstruction is frequently detected after pulmonary artery banding for the following reasons: 1) initially latent subaortic stenosis may become apparent after the placement of a pulmonary artery band causes an increase in the systemic to pulmonary flow ratio; 2) the natural course of orifice size of the ventricular septal defect is to diminish with time (22); and 3) some investigators (25) believe that placement of a pulmonary artery band leads to increased ventricular hypertrophy, thereby accelerating the process of ventricular septal defect size diminution and subaortic obstruction.

We believe that for the patients described in our series, pulmonary artery banding early in infancy may be detrimental to long-term outcome. In the setting of obvious obstruction to aortic outflow, such banding may lead to ventricular failure secondary to severe afterload. In infants without obvious subaortic obstruction, but with arch abnormalities, a predisposition toward the development of subaortic obstruction has been shown (26). Pulmonary artery banding, although a solution to the immediate problem of pulmonary overcirculation, may lead to harmful changes in ventricular compliance. Patients with such banding are therefore at high risk for increased morbidity and mortality during a subsequent Fontan surgical procedure. As do others (27), we recommend that, instead of pulmonary artery banding, a pulmonary artery to aorta anastomosis with placement of a shunt be performed in infancy as part of a staged surgical approach to definitive palliation for these complex lesions.

Management strategy for infants with aortic outflow obstruction. In complex lesions of functionally single ventricle where the potential exists for subaortic obstruction despite the absence of a significant gradient, careful anatomic delineation of aortic anulus and ventricular septal defect size, as well as the presence or absence of aortic arch obstruction, should be noted. In infants with diminished cardiac output, early diagnosis, rapid correction of metabolic imbalances and institution of propranolol infusion for ducal patency are crucial. A balance between pulmonary and systemic vascular resistances must be maintained. The scenario of high pulmonary flow (Pp h >40 mm Hg) at the expense of systemic perfusion with the development of metabolic acidosis should be avoided by the reduction of inspired oxygen to 21%. Hyperventilation and pulmonary overcirculation may be avoided with the use of paralysis and controlled mechanical ventilation (9).

Survival after initial palliation. Similar to the experience with infants after initial palliation for hypoplastic left heart syndrome (9), early mortality in our patients was proportionately high: 82% of deaths occurred within the 1st 30 days and half of these occurred within the 1st 24 h. Almost all of the early deaths are due to a rapid and sudden cardiovascular collapse that we believe is secondary to sudden changes in the pulmonary to systemic vascular resistance ratio.

Of the variables analyzed, only aortic arch anatomy appears to have some correlation with early survival, although the numbers are too small to achieve statistical significance. Eleven (32%) of 34 infants with arch anomalies died within the 1st 30 days after operation, with 3 additional deaths occurring after 30 days and before the next surgical stage was reached; 3 (19%) of 16 infants without arch anomalies died early. with no subsequent mortality occurring until the next surgical stage. Arch obstruction was not a frequent cause of death in the group with arch anomalies; the only patient with an arch-related cause of death had a complex anomaly consisting of double aortic arch with right cervical arch and interruption of the left arch. Arch dissection was detected in this patient on the 2nd day after operation and, despite an emergency attempt at repair, the patient died. The three late deaths in this group were also unrelated to the aortic arch: one was due to sepsis and two were sudden and unexplained.

Superior cavopulmonary anastomosis. Chronic volume overload seen in a functionally single ventricle after placement of a systemic to pulmonary artery shunt leads to compensatory changes in the myocardium that are detrimen-
tal at the time of the Fontan procedure. Ventricular muscle mass is chronically increased in response to pumping both the pulmonary and systemic flows. Increased ventricular muscle mass is reported to be a high risk factor in patients undergoing the Fontan procedure (28). In addition, at the time of the Fontan procedure, pulmonary and systemic circulations are separated and geometric changes occur because of the sudden mismatch between ventricular volume and mass. End-diastolic volume suddenly diminishes and therefore the ratio of ventricular wall thickness to end-diastolic volume increases. This results in increased ventricular stiffness, a decreased driving gradient across the pulmonary and systemic bed and diminished cardiac output in the early postoperative period.

Since May 1989, we have reduced the volume overload before the Fontan procedure in an attempt to avoid this phenomenon by takedown of the systemic to pulmonary shunt and anastomosis of the superior vena cava to the augmented pulmonary arteries at an early age (6 to 12 months). In the present series, this technique was used in 13 patients without deaths and the Fontan procedure was completed in 4 patients 6 months later by patch baffling inferior vena cava flow to the superior vena cava-pulmonary artery junction. Whether the interposition of this hemi-Fontan will significantly improve survival after completion of the Fontan procedure is not yet known.

Obstruction to aortic outflow with normal mitral valve and normal left ventricle. Within the heterogeneous group of lesions with obstruction to aortic outflow, a small subset of patients exists with normal left ventricular dimensions, a normal size mitral valve and a large unrestrictive ventricular septal defect, usually of the malalignment type (5–8). These patients are suitable candidates for utilization of the left ventricle as the systemic pumping chamber. In our series, eight patients were suitable for a biventricular repair in conjunction with a pulmonary artery to aorta anastomosis. Although successful performance of a primary biventricular repair in infancy has been reported (29,30), all three of our infants who underwent primary biventricular repair died (two early, one late). All five infants who underwent initial pulmonary artery to aorta anastomosis and later takedown of systemic to pulmonary shunt with performance of biventricular repair have survived and are doing well.

Conclusions. Obstruction of aortic outflow may be seen in complex lesions other than the hypoplastic left heart syndrome. Performance of a pulmonary artery to aorta anastomosis with placement of a systemic to pulmonary shunt and arch augmentation as necessary can be successfully employed in infancy and results in a hemodynamic status favorable for a subsequent Fontan procedure. Survival after a pulmonary artery to aorta anastomosis is equal among various anatomic groups of ventriculoarterial alignment, whereas arch anatomy appears to influence survival and more deaths are seen in infants with aortic arch abnormalities. A pulmonary artery to aorta anastomosis can be successfully utilized as an initial palliative procedure with later performance of a biventricular repair in infants with obstruction to aortic outflow, ventricular septal defect and normal size left ventricle and normal mitral valve. We speculate that by the interposition of a bidirectional cavopulmonary anastomosis (hemi-Fontan procedure) before the Fontan procedure is performed, early reduction of volume overload will be achieved, thereby preventing detrimental ventricular geometric changes.

References