Biventricular Repair Approach in Ducto-Dependent Neonates With Hypoplastic but Morphologically Normal Left Ventricle

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Objective

Increased afterload and multilevel LV obstruction is constant. We assumed that restoration of normal loading conditions by relief of LV obstructions promotes its growth, provided that part of the cardiac output was preoperatively supported by the LV, whatever the echocardiographic indexes.

Background

Whether to perform uni- or biventricular repair in ducto dependent neonates with hypoplastic but morphologically normal LV (hypoplastic left heart syndrome classes II & III) remains unanswered. Echocardiographic criteria have been proposed for surgical decision.

Methods

Twenty ducto dependent neonates presented with this anomaly. All had aortic coarctation associated to multilevel LV obstruction. Preoperative echocardiographic assessment showed: mean EDLVV of 12.4 ± 3.03 ml/m² and mean Rhodes score of −1.73 ± 0.8. Surgery consisted in relief of LV outflow tract obstruction by coarctation repair in all associated to aortic commissurotomy in one and ASD closure in 2.

Results

There were 3 early and 2 late deaths. Failure of biventricular repair and LV growth was obvious in patients with severe anatomic mitral stenosis. The other demonstrated growth of the left heart. At hospital discharge the EDLVV was 19.4 ± 3.12 ml/m² (p = 0.0001) and the Rhodes score was −0.38 ± 1.01 (p = 0.0003). Actuarial survival and freedom from reoperation rates at 5 years were 72.5% and 46%, respectively.

Conclusions

Biventricular repair can be proposed to ducto dependent neonates with hypoplastic but morphologically normal LV provided that all anatomical causes of LV obstruction can be relieved. Secondary growth of the left heart then occurs; however, the reoperation rate is high.

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Manuscript received February 19, 1998; revised manuscript received September 18, 1998, accepted November 20, 1998.

Journal of the American College of Cardiology Vol. 33, No. 3, 1999
© 1999 by the American College of Cardiology ISSN 0735-1097/99/$20.00 Published by Elsevier Science Inc. PII S0735-1097(98)00636-6

Hypoplastic left heart can include a spectrum of underdeveloped left-sided heart structures and aortic outflow tract. At the severe end of this spectrum there is the hypoplastic left heart syndrome with almost total absence of left ventricle and at the other less severe extremity there is the left ventricular hypoplasia but with morphologically normal left heart. After birth, due to the closure of ductus arteriosus, these patients disclose a severe heart failure which is reversible with the use of Prostaglandins E1. Without intervention, evolution is almost always fatal in the first week. The surgical treatment of hypoplastic left heart syndrome and variants has been well codified in the recent years and includes multiple step procedures with still improving early and long term results (1,2).

Patients born with the less severe form of hypoplastic left heart still present controversial surgical management. After birth whether to determine if the left ventricle will be able to sustain a systemic output without a ductus is still unanswered. In an attempt to make a classification, Kirklin (3) proposed that this should be considered to represent the hypoplastic left heart syndrome. At one end of this syndrome is the aortic atresia with almost complete absence of left ventricular cavity (HLHS class IV) and at the other end are those having isolated anomaly of the left ventricle from a list of six, including congenital mitral valve disease, left ventricular hypoplasia with concordant ventriculoarterial connection, subvalvar or valvar or supravalvar aortic stenosis or aortic arch hypoplasia, interrupted aortic arch, or coarctation (HLHS class I). Those having two of the six congenital anomalies affecting the left ventricular outflow tract are classified HLHS class II. In between, are those patients with multilevel left ventricular obstructions and hypoplasia of the left ventricle (HLHS class III). Definition of hypoplastic left heart syndrome class III includes the
presence of more than two of the six anomalies, or two with coexisting left ventricular or ascending aortic or aortic arch hypoplasia. Although there is no controversy with regard to the medico surgical management for HLHS class I and IV, in some patients with HLHS class II, particularly those with severe left ventricular hypoplasia, the medico surgical management remains a difficult problem because of several undefined criteria and this lack of decision making is even more pronounced in patients with HLHS class III. The extent of left ventricular hypoplasia has been defined as the association of a mitral valve area less than 4.75 cm²/m², left ventricular inflow dimension less than 25 mm, an indexed end diastolic left ventricular volume below 20 ml/m², a ratio between the apex to base dimension of the left ventricle and that of the right ventricle of less than 0.8 or a transverse cavitary and aortic annular dimension of 6 mm or less (3).

Biventricular repair presents obvious advantages over univentricular type of repair, but it might carry a higher operative risk for early mortality. On the other hand, if univentricular heart repair option is decided, the cumulative risks of several operations and the natural evolution of a single ventricle circulation might be considered, even in the case of a “perfect” candidate. Echocardiographic criteria have been proposed to select patients in whom the small left heart will not be able to sustain a systemic output postoperatively. In neonates with critical aortic stenosis, Rhodes et al. (4) proposed that a patient will be able to sustain systemic output if he presents with a score > −0.35 and three of the following: (i) long axis ratio >0.8, (ii) aortic root diameter indexed to BSA >3.5 cm/m², and (iii) mitral valve area indexed to BSA >4.75 cm²/m² and left ventricular mass indexed to BSA >35 g/m². Other authors proposed additional criteria precluding biventricular repair. They include a mitral annulus diameter below 9 mm (5), a left to right ventricle end diastolic diameter ratio below 0.33 (6) and a heart apex made by the right ventricle (7).

Recent reports (8,9) have shown that these criteria are not as strict as they seem to be. Minnich et al. (9) reported seven infants with small left heart structures, all presenting with a Rhodes score under the feasibility of biventricular repair. After medical intensive therapy for weeks, echocardiographic measurements of left heart structures and Rhodes score had increased within ranges allowing biventricular repair.

Our experience with biventricular repair in those patients was first attempted at a period when the univentricular option was not as popular as it is nowadays. The good outcome of these initial patients prompted us then to develop an approach in which biventricular repair should be favored. Rather to reason in terms of anatomic lesions associated with small left heart, we preferred a more physiological approach in which one could point out an anatomic lesion as a factor for left ventricular underdevelopment. We could therefore distinguish three main types of physiological abnormalities: (i) insufficient preload to the left ventricle (HLHS Class I), (ii) an increased afterload to the left ventricle (HLHS Class II), and (iii) mixed physiological anomalies (HLHS Class III). In the first category, the situation is somehow different because the patients were generally older and not ducto dependent. On the other hand, it has been previously demonstrated (10,11) that reestablishment of a near normal preload to the left ventricle restores a normal left ventricular output and size.

As a whole, it was considered that surgery has as a main objective to normalize the loading conditions of the left ventricle by increasing the preload and/or decreasing the afterload. In the latter group of mixed physiopathological conditions, it was felt, however, that normalization of loading conditions should be performed stepwise and not in a single stage operation.

Beside the potential operative mortality of biventricular repair, growth of the left heart after initial repair is another important issue. Is the left heart able to grow harmoniously after initial surgery parallel with body growth without reoperation or pharmacological support?

We report herein our experience with biventricular surgical repair in patients with severe HLHS Class II and HLHS class III.

PATIENTS AND METHODS

From January 1984 to May 1998, 20 ducto dependent neonates, eight with a severe hypoplastic left heart syndrome class II and 12 with HLHS class III were referred to our Institution for surgery. Since 1990, biventricular repair was decided versus univentricular option or heart transplantation after staff and parents discussion. During this period, only two patients with the same anatomy underwent a Norwood stage I procedure.

Inclusion criteria. All patients with hypoplastic left heart syndrome classes II and III were retrospectively included in this series provided that the echocardiographic videotape was fully exploitable. The inclusion criteria were as follows: 1) aortic arch obstruction and left ventricular hypoplasia, 2) the degree of left ventricular hypoplasia accepted for including patients was based on an end diastolic left ventricular volume less than 20 ml/m² with a mitral valve area index of less than 4.75 cm²/m² and aortic root diameter less than 3.5 cm/m², 3) patent aortic and mitral valves with an anterograde flow, 4) ductal dependency without retrograde flow in the ascending aorta but either right to left or bidirectional ductal shunt, 5) intact ventricular septum, and 6) Rhodes score of less than −0.35.

Preoperative echocardiographic assessment. All the patients underwent serial echocardiographic studies to assess the intra- and extracardiac anatomy and to perform measurements. Aortic annulus size and aortic root were measured from a parasternal long-axis view. Dimensions of the transverse arch and of the aortic isthmus were obtained from suprasternal images. The mitral valve anteroposterior diam-
Atrioventricular valve areas were then calculated with the formula for an ellipse. Left ventricular volumes were calculated as previously described (4). All the measurements were indexed to BSA. Left ventricular end diastolic diameter was obtained from parasternal long-axis view and lateral diameter from the apical four chamber views at end diastole. Aortic stenosis was diagnosed in 10 fetuses allowing delivery close to the surgical unit. Prior to surgery, 68% were already intubated and supported by artificial ventilation and inotropic support. Ductal patency was maintained in all by PGE1 infusion, one having a spontaneously patent ductus arteriosus. The details of anatomic lesions and echocardiographic assessments are listed in Table 1. Briefly, all had isthmic coarctation of the aorta associated with an hypoplastic transverse arch. One with associated coarctation had congenital aortic stenosis. In addition, all presented various types of left-sided multilevel obstruction and important left to right atrial shunting by the way of an atrial septal defect either primum or secundum. As well, none demonstrated retrograde ductal flow in the ascending aorta.

**Echocardiographic sizes of the left heart (Table 1).** All the patients presented with hypoplastic but morphologically normal left ventricle. The left ventricle formed the apex in eight patients whereas in 13, the apex was formed by the right ventricle. Both mitral and aortic valves were patent and normal left ventricle. The left ventricle formed the apex in eight patients whereas in 13, the apex was formed by the right ventricle. Both mitral and aortic valves were patent and

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**Table 1. Patients Population**

<table>
<thead>
<tr>
<th>Pts</th>
<th>Age (Days)</th>
<th>Weight (kg)</th>
<th>Diagnosis</th>
<th>HLHS Class</th>
<th>Mitral Diameter (Z-value)</th>
<th>Aortic Diameter (Z-value)</th>
<th>LVEDV (ml/m2)</th>
<th>Rhodes Score</th>
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<td>12.5</td>
<td>-1.38</td>
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CoA = Coarctation; ASD = atrial septal defect; pAPVR = partial anomalous pulmonary venous return; pAVC = partial atrioventricular canal; AS = aortic stenosis; PFO = patent foramen ovale; MS = mitral stenosis.
none of them presented with retrograde flow in the ascending aorta from the ductus arteriosus. The ductal shunt was right to left in five patients and bidirectional in 16. The mean mitral annulus diameter was: 7.5 ± 1.2 mm (mean Z-value to normal: −3.32 ± 1.6). The mean aortic annulus diameter was: 5.4 ± 1.4 mm (mean Z-value to normal: −7.2 ± 2.8). The mean end diastolic left ventricular diameter was 12.65 ± 1.4 mm. The mean left ventricular end diastolic volume was: 12.4 ± 3 ml/m² and the mean Rhodes score was: −1.73 ± 0.8.

Surgical approach. The first surgical procedure consisted in all patients in treatment of the anatomical cause for increased afterload. Seventeen patients underwent repair of the aortic coarctation by extended end to end anastomosis through left thoracotomy. In three patients, coarctation repair was undertaken through midline sternotomy and was associated in two cases to closure of ASD and aortic commissurotomy in one. Inadequacy of the left ventricle to sustain a systemic output and failure of left ventricular development were determined by postoperative death, early conversion to univentricular circulation and absence of left ventricular growth at repeated echocardiography.

Follow-up. Repeated echocardiographic measurements were performed before hospital discharge to assess if any growth of the left heart had already occurred. Follow up was achieved in 100% of survivors by telephone calls and correspondence with the referring pediatric cardiologists. Gross information about the growth of the left heart structures was particularly noted at each visit and more accurately if reoperation was required. The median follow-up was: 53 months (Ranges: 3–159 months).

Statistical analysis. The relation between failure of biventricular repair and survival to age at presentation, body surface area and each of the echocardiographic measurements was examined by stepwise logistic regression. Echocardiographic indices of left heart sizes were compared prior to and after surgery by the Student t test.

Time related events were examined by Kaplan-Meyer actuarial methods. Ratios were expressed with 70% confidence limits (CL).

RESULTS

Postoperative course. The median stay on ventilator was six days, ranging from 2 to 75 days. Three patients undergoing aortic arch repair through midsternotomy, cardiopulmonary bypass and circulatory arrest had delayed sternal closure. Following surgery all the patients disclosed normal aortic saturation and only two following aortic arch repair and ASD closure had a metabolic acidosis. Two other patients were not weanable from ventilatory support and were reoperated on for fenestrated ASD closure within two weeks of the initial aortic arch repair. In another case, although this patient had an initial uneventful post operative course and could be extubated within 4 days following surgery, he developed a low cardiac output one week after discharge from the ICU and underwent repeated cardiac catheterization which demonstrated low cardiac output, severe mitral stenosis with a mean gradient of 8 mm Hg and subaortic tunnel with a peak systolic gradient of 50 mm Hg. He was unsuccessfully reoperated on for mitral valve repair and Ross procedure. The median length of hospital stay was 15 days (Ranges: 10 to 75 days).

Mortality. There were three early deaths due to inadequacy of the left ventricle to sustain a systemic output. Two of them underwent single stage complete repair of aortic coarctation and closure of ASD. In these two patients, closure of ASD was felt to improve preloading of the left ventricle and to promote its growth. However one of them had parachute mitral stenosis and died one month later after an attempt at mitral valve repair. The other died one month after surgery in a low cardiac output and end stage multiple organ failure. A 2 kg patient with Shone syndrome had coarctation repair. Few days after extubation, he developed a low cardiac output. Repeated echocardiography and angiography demonstrated a severe parachute mitral stenosis associated to severe subaortic tunnel. He was unsuccessfully reoperated on for mitral valve repair and Ross procedure.

One patient died two months after initial surgery. He had isthmic coarctation associated with an ostium primum and parachute mitral valve stenosis. Although coarctation repair was initially well tolerated, the left ventricle did not demonstrate any growth and the patient suffered severe pulmonary hypertension related to the mitral stenosis. He was unsuccessfully converted to univentricular circulation by the age of two months and a half. None of the recorded echocardiographic indices showed a significant risk factor for inadequacy of the left ventricle to sustain a systemic output postoperatively.

Reoperations. Four patients required an early reoperation during the same hospital stay. They were previously described. Briefly, two not weanable from ventilatory support underwent fenestrated closure of a large atrial septal defect with an uneventful postoperative course. The others with residual inlet and outlet LV obstruction underwent isolated mitral valve repair once and the other had mitral valve repair associated to a Ross procedure. Both died after reoperation.

Eight patients underwent 12 late reoperations within a mean delay of 30 ± 26 months postoperatively with one death. The different reoperations are listed in Table 2. At time of late reoperation, the size of left heart structures reached normal values.

Growth of left heart structures (Fig. 1). Within a median of 15 days following initial surgery, all the echocardiographic measurements of left heart structures demonstrated a significant increase after coarctation repair. The mitral valve annulus grew from 7.5 ± 1.2 mm to 9 ± 1.3 mm (p = 0.0002). The aortic valve annulus grew from 5.4 ± 1.4 mm to
to 6.53 ± 1.5 mm (p = 0.0002). The end diastolic left ventricular volume increased from 12.65 ± 1.4 ml/m² to 18.93 ± 3.15 ml/m² (p = 0.0001). The Rhodes score was recalculated immediately before hospital discharge; it was 2 ± 1.73 ± 0.8 preoperatively and 2 ± 0.51 ± 1 before hospital discharge (p = 0.0003).

Patients who had echocardiographic demonstration of anatomic mitral stenosis disclosed a different evolution. Seven patients had an LV inlet obstruction with a mean echocardiographic gradient of 6 mm Hg due to parachute mitral valve in six and to supravitral ring in one. In this cohort, the left heart structures did not grow enough to support a systemic output. Three patients died after early reoperation and two required a reoperation within three months of the initial coarctectomy to trigger growth of the LV. Finally, one patient was reoperated on twice 40 months later for mitral valve replacement but suffered soon after reoperation mitral prosthetic thrombosis. He died after the second emergent reoperation. At the time of reoperation, he had a normal sized left ventricle. Although mortality was higher in this group of patients than in those who did not have anatomic mitral stenosis, it did not reach statistical significance, probably due to the small number of patients.

**Follow-up.** All the survivors were followed by their referring pediatric cardiologists. The median follow-up was 53 months (Ranges: 3–159 months). All the survivors demonstrated a normalization of left heart size without pulmonary hypertension at the last echocardiographic control and were medication free.

The actuarial survival and freedom from reoperation rates at five years postoperatively are illustrated in Figure 2.

**DISCUSSION**

Hypoplastic but morphologic normal left ventricle is a clinical entity which remains difficult to define. It generally associates aortic coarctation and relative hypoplasia of the left ventricle with small mitral and aortic valves. Kirklin's classification (3) presents the advantage of unifying different anatomical entities. It does not, however, take into account the physiologic events underlying the various forms of hypoplastic left ventricle. In addition, it is sometimes difficult to distinguish between HLHS class II and HLHS class III. Many investigators have tried to determine those patients who will survive a two-ventricle repair and those who must be treated as having hypoplastic left heart syndrome. Initial studies were based on echocardiographic measurements of the end diastolic left ventricular diameter (EDLVD) (16) or indexed end diastolic left ventricular volume (EDLVV) (17,18). An EDLVD below 13 mm or an EDLVV below 20 ml/m² were strong predictors for poor outcome after biventricular repair. More recently, Rhodes...
delineated a score to identify candidates for one or two ventricle repair (4) for patients suffering a congenital aortic stenosis. Finally the presence or absence of an endocardial fibroelastosis was also found to be a discriminant factor for survival after two ventricle repair (19,20). In the present series, all the patients had an EDLVV below 20 ml/m2 and a Rhodes score below the feasibility of biventricular repair. However, all presented with aortic coarctation and hypoplastic transverse aortic arch associated with various degree of multilevel left ventricular obstruction and only one had associated congenital aortic stenosis. Also none presented endocardial fibroelastosis. They were all ducto dependent at initial presentation but none had a retrograde flow from the ductus in the ascending aorta, indicating that a substantial portion of the systemic output was provided by the left ventricle. The mitral valve, although stenotic and/or hypoplastic in several patients, always demonstrated a forward flow despite an important left to right atrial shunt.

In addition to the poor echocardiographic indices, biventricular repair was decided in these patients for two main reasons. First, the circulatory conditions were very similar to those of fetal circulatory patterns. Namely, the entire cardiac output was maintained by both left and right ventricles, the latter through the patent ductus arteriosus. During fetal life, due to these circulatory patterns, the right ventricle is dominant while the left ventricle remains smaller. Following birth, with the fall in pulmonary vascular resistances, closure of the ductus arteriosus and closure of the foramen ovale, there is an increased pulmonary venous return which promotes growth of the left ventricle. In cases of small left ventricle with multistaged left ventricular obstruction, use of PGE1 allows persistence of fetal circulatory patterns. However, because there is a fall in pulmonary vascular resistances with increase in pulmonary venous return but still a small left ventricle, the foramen ovale shunts left to right until restoration of an unobstructed left ventricle. Therefore restoration of an unobstructed left ventricular inflow or outflow tracts by repair of coarctation with ductus division corresponds to the physiologic events occurring after birth in normal hearts. Progressive adaptation of the left ventricle to these new loading conditions then occurs and the foramen ovale generally closes spontaneously. The presence of an inflow left ventricular obstruction by the way of virtual hypoplasia of the mitral valve slows down the growth of the left ventricular cavity by a mechanism of autoregulation probably dependent on the left ventricular compliance. Indeed, in these patients repeated echodoppler studies were able to demonstrate a left to right shunt at the atrial level immediately after coarctation repair which became bidirectional within two weeks. In cases of anatomical mitral stenosis, although left ventricular filling was improved following coarctation repair, left ventricular output remained inadequate to sustain a systemic output. Direct surgical approach to repair stenotic mitral valve was unsuccessful in 50% of cases (2/4) as was conversion to univentricular circulation in 100% (1/1). Second, according to the concept that intrauterine flow patterns are important trophic stimulus for the normal development of cardiac structures (21), any lesion which reduces blood flow through the different components of a normal left ventricle would be expected to lead to underdevelopment of downstream structures; therefore, this previous scenario is made possible provided that the different levels of obstruction to the left ventricle are treated. In this series the lowest mitral valve annulus diameter of 6 mm was associated with survival and rapid growth was demonstrated. On the other hand patients with a larger diameter of the mitral valve but with anatomical stenosis due to parachute valve was not associated with survival. Left ventricular outlet obstruction can occur at the subaortic region, at the aortic anulus or valvar level or at the aortic isthmus either isolated or in association. Whether an
hypoplastic aortic annulus can sustain a systemic output is still under debate. In the present series the lowest aortic annulus diameter associated with survival was 4 mm. All the nonsurvivors had aortic annulus sizes ranging between 4 and 6 mm. Recent application of the Ross–Konno procedure in neonates should be an attractive solution for these patients (22); it was, however, unsuccessfully applied in one patient with associated mitral stenosis. The last cause of anatomical left ventricular restriction is the endocardial fibroelastosis. Although none of the patients of this series presented this lesion, it has been found to be an important risk factor to the outcome because it impairs left ventricular compliance. Finally, we believe that the left ventricular muscle without endocardial fibroelastosis is able to stretch relative to the quantity of cardiac output flowing through whatever the initial degree of left ventricular underdevelopment. True obstruction at the mitral valve annulus and apparatus or at the aortic valve annulus and subaortic region are the limiting factors to left ventricular adaptation to increase of cardiac output if still stenotic. In case of residual mitral obstruction, the left ventricle cannot grow or stretch and leads to important left to right atrial shunt and pulmonary hypertension. In case of severe residual aortic obstruction, there is distension of the left ventricle with eventual myocardial failure.

Based on this hypothesis, all the patients from the present series underwent biventricular repair approach. Overall failure of this approach was 20% which compares well with series of stage I procedure in this subset.

Altogether, indications for biventricular repair does not rely only on the echocardiographic measurements and deviations from normal values but also on a physiologic and clinical assessment of the intracardiac flow patterns and on the surgical aptitude to relieve all obstruction to the left ventricle. The most critical patient from this series was a neonate with aortic coarctation only, end diastolic left ventricular diameter was 12 mm, EDLVV was 4 ml/m2 and Rhodes score was −2.47. The mitral valve diameter was 6 mm, the aortic valve and subaortic diameters were 4.5 mm. This patient had, however, already at antenatal diagnosis and at postnatal examination a forward flow through the mitral and aortic valves and no retrograde flow from the ductus to the ascending aorta. He underwent coarctation repair and demonstrated a rapid growth of left heart. On the other hand, patients with higher scores and echocardiographic indices failed when this approach was used. Analysis of these failures pointed out several important findings. Three out of four failures were related to residual severe mitral stenosis. Although there was a preoperative echocardiographic documentation of this lesion, it remained difficult to assess the degree of severity of the mitral lesion in presence of an ASD shunting left to right. It was therefore assumed that, since part of the cardiac output flowed through the mitral valve preoperatively, it should be able to sustain a systemic output following coarctation repair and ductus division. Interesting enough all the patients with mitral stenosis who were not able to sustain biventricular repair preoperatively presented an exclusively right to left shunt at the ductal level which indeed could reflect the degree of mitral stenosis severity. Another newborn died after repair of coarctation and closure of atrial septal defect in a single stage operative procedure. The obligatory forward flow through the mitral valve after ASD closure has probably caused a left ventricular distension while compliance was still inadequate.

From these observations, it can be proposed that in cases where cardiac output is partially dependent of the preoperative ductal patency, the left ventricle does not participate in the entire cardiac output. Division of the ductus associated with a free patent left ventricular inflow and outflow tracts allows reestablishment of circulation in series which can progressively adapt the left ventricular output by shunting through the foramen ovale. Therefore closure of any atrial shunt forcing the entire output through the left ventricle would be expected to result in an LV overload and distension. Neonatal LV already functioning at the plateau level of Starling’s curve, any volume overload will end up with severe LV dysfunction and eventually patient’s death. When the left ventricle has demonstrated its ability to adapt to the new loading conditions, then closure of residual shunts might be performed in order to improve the systemic output as it was necessary in two patients of this series. In presence of anatomic mitral stenosis, the severity of the latter indicating either direct surgical approach or univentricular approach can be evaluated by the direction of the ductal shunt. In these conditions we are, at present time, still unable to assess preoperatively which type and degree of mitral stenosis that will be able to sustain a near normal forward flow and systemic output and which will not.

From the data of this series, the aptitude of the left ventricle to grow after reestablishment of a near normal physiology is remarkable. However, at present time, there is no evidence that growth of the left heart results in cardiomyocytes multiplication and/or elongation or a more global left ventricle remodeling (23).

The rate of reoperation was 61.9% at five years. The main causes of reoperation were reappearance of an obstructive lesion to the left ventricle either at the inlet or at the outlet. Therefore, although the left ventricle is able to demonstrate a harmonious growth, it is not free from reappearance of stenotic lesion requiring reoperation.

In conclusion, ducto dependent neonates born with an hypoplastic but morphologically normal left ventricle (HLHS classes II-III) can be candidates for biventricular repair provided restoration of an unobstructed left ventricular blood stream. Initial correction by coarctation repair and ductus division is generally sufficient to restore a near normal cardiac output through the left ventricle. The residual patent foramen ovale is a useful discharge for autoregulation of the cardiac output during the first postoperative days. The single limiting factors to biventricular repair in this subset of patients are the semirigid structures
merging the left ventricle, namely the mitral and the aortic regions. If they are not amenable to surgical patency, the univentricular option should be preferred.

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REFERENCES