

Borderline hypoplasia of the left ventricle in neonates: Insights for decision-making from functional assessment with magnetic resonance imaging

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Objectives: We sought to compare the usefulness of echocardiography and magnetic resonance imaging in neonates with a borderline small left ventricle.

Methods: The preoperative magnetic resonance and echocardiography studies of 20 consecutive patients (mean age 10 ± 9 days) undergoing magnetic resonance imaging were analyzed. The diagnoses were aortic stenosis ($n = 3$), hypoplastic left heart complex ($n = 12$), and unbalanced atrioventricular septal defect ($n = 5$). The magnetic resonance imaging protocol included ventricular volumetry, flow measurements, and angiography. Potential left ventricular volumes, assuming an ideal geometric shape, were calculated by mathematically “unfolding” the compressed left ventricle.

Results: Left ventricular end-diastolic volume was 16.0 ± 7.0 mL/m² of body surface area by echocardiography and 33.5 ± 15.5 mL/m² by magnetic resonance imaging. Echocardiography consistently underestimated left ventricular volume and did not correlate with magnetic resonance. Of all echocardiographic parameters, mitral valve z-score was the best predictor of left ventricular end-diastolic volume by magnetic resonance ($r = 0.77$; $P = .02$). The average potential volume increase was 8.8% for aortic stenosis, 35.0% for atrioventricular septal defect and 23.0% for hypoplastic left heart complex patients. Aortic valve diameter did not correlate with flow volume in the ascending aorta. Sixteen (80%) of 20 patients underwent biventricular repair, without early mortality. Of these, only 5 (31.3%) had a preoperative left ventricular end-diastolic volume of more than 20 mL/m² by echocardiography.

Conclusions: Magnetic resonance imaging is feasible in neonates with borderline left ventricular hypoplasia. Echocardiography does not accurately measure left ventricular hypoplasia in these patients and may unfairly preclude some patients from a biventricular repair in whom magnetic resonance is reassuring.

Almost 50 years ago Noonan and Nadas¹ coined the term “hypoplastic left heart syndrome,” referring to a spectrum of cardiac anomalies characterized by varying degrees of underdevelopment of the left heart and aortic arch. At the severe end of the spectrum, the aortic valve (AV) is atretic, the mitral valve (MV) is severely hypoplastic, and the diminutive left ventricle (LV) shows endocardial fibroelastosis. At the mild end, hearts with small, but not intrinsically stenotic AVs and MVs and without endocardial fibroelastosis have been termed “hypoplastic left heart complex (HLHC).”² That spectrum, however, is dichotomized by the binary surgical decisions of biventricular (BV) versus univentricular (UV) repair made early in life. The consequences of the wrong decision, if made toward a BV repair, are often detri-

mental in the short term, whereas unnecessary selection toward a UV course may have adverse long-term functional outcomes.

We recognized the shortcomings of echocardiography, which is the main diagnostic tool in assessing the adequacy of the LV to support the systemic circulation after the BV repair. We hypothesized that preoperative functional magnetic resonance imaging (MRI) is feasible in neonates with borderline hypoplasia of the LV and would aid in the decision between UV and BV repair.

This report summarizes our initial experience in 20 patients with a small LV as measured by echocardiography who underwent MRI before their first intervention.

METHODS

Between March 2003 and August 2006, 20 consecutive patients with borderline hypoplasia of the LV and in whom there was controversy regarding the decision between BV and UV repair underwent a preoperative cardiac MRI. All patients had an LV end-diastolic volume (LVEDV) of less than 30 mL/m² of body surface area by the Simpson volumetric analysis and none of them had obvious cardiovascular anatomy demonstrated on echo that precluded a BV approach. There were three distinct groups: (1) patients with intrinsic critical AV stenosis (AS, $n = 3$), (2) patients with structurally normal MV and AV and hypoplastic arches, grouped as HLHC, with or without coarctation ($n = 12$), and (3) patients with unbalanced atrioventricular septal defects (AVSD, $n = 5$). All studies were

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Received for publication Dec 25, 2007; revisions received March 10, 2008; accepted for publication April 13, 2008.

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J Thorac Cardiovasc Surg 2008;136:1429-36
0022-5223/\$34.00

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doi:10.1016/j.jtcvs.2008.04.027

Abbreviations and Acronyms

- AAO = ascending aorta
- AS = aortic valve stenosis
- AV = aortic valve
- AVSD = atrioventricular septal defect
- BV = biventricular
- DAO = descending aorta
- HLHC = hypoplastic left heart complex
- LV = left ventricle
- LVEDd = left ventricular end-diastolic diameter
- LVEDV = left ventricular end-diastolic volume
- MRI = magnetic resonance imaging
- MV = mitral valve
- PCMRI = phase-contrast magnetic resonance imaging
- RV = right ventricle
- UV = univentricular

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performed on a 1.5-Tesla scanner (GE signa CV/i; General Electric Medical Systems, Milwaukee, Wis) using a head coil. All but one patient were intubated during the study. Ventricular volumes and flow volumes were calculated on a commercially available work station (Mass Analysis and CV Flow; MEDIS Medical Imaging Systems, BV, Leiden, The Netherlands). In patients with AVSD, LV volumes were measured by MRI and echocardiography by virtually extending the deficient interventricular septum to the base of the heart.

The MRI protocol consisted of cine imaging in axial, 2-chamber, 4-chamber, and short-axis planes, through-plane phase-contrast imaging (PCMRI) of the main, right, and left pulmonary arteries, ascending aorta (AAO) at the level of the right pulmonary artery, descending aorta (DAO) at the diaphragm, patent ductus arteriosus, superior vena cava, and atrioventricular valves, and contrast-enhanced MRI angiography.

True and Potential LV Volumes

The short-axis cine imaging for ventricular volumetry (Figure 1) was performed by using a segmented spoiled gradient refocused echo sequence with minimum echo and repetition times, flip angle 20°, bandwidth 31.25 kHz, number of excitations of 2, slice thickness 3.6 to 5.0 mm, number of slices 5 to 11 without gap, minimum field of view (160–200 mm), matrix 256 × 160, 20 reconstructed phases per cardiac cycle.

Phoon and Silverman³ introduced the model of observed versus potential preoperative volume to highlight the concept that the LV may be underfilled and compressed by the volume-loaded right ventricle (RV), leading to an underestimation of its potential postoperative volume (Figure 2). Potential LV volume was defined as the expected volume when the crescentic LV is mathematically converted to an ellipsoid chamber. This is the expected shape after effective relief of RV overload and adequate LV filling. We applied this method to MRI volumetry. The endocardial circumference (C) of the LV cavity was measured in end-diastole in each short-axis plane. Assuming that C would be maintained after conversion of the crescentic to a round configuration, the potential LV area in each short-axis plane (A_{pot}) and potential LV volume (V_{pot}) were calculated by using the following formulas:

$$A_{pot} = C^2/4\pi \tag{1}$$

$$V_{pot} = \sum A_{pot \ 1-n} \times \text{slice thickness} \tag{2}$$

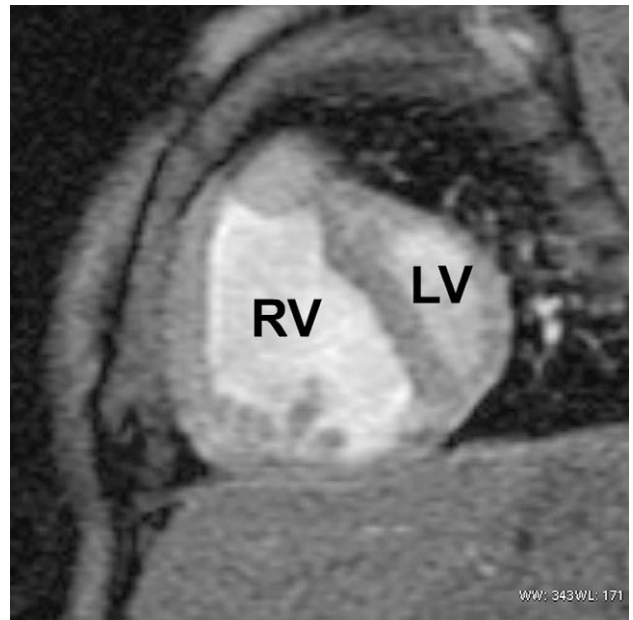


FIGURE 1. Short-axis slice at end-diastole in a patient with unbalanced atrioventricular septal defect. The left ventricle (LV) is compressed by the right (RV). The left atrioventricular valve is suspended by a single papillary muscle.

Blood Flow

Imaging parameters for PCMRI were as follows: Minimum echo and repetition times, flip angle 20°, bandwidth 31.25 kHz, 1 to 2 k-space lines per segment, number of excitations 2, slice thickness 4 mm, minimum field of view (160–200 mm), matrix 256 × 160, velocity encoding 150 cm/s, 20 reconstructed phases per cardiac cycle. Pulmonary blood flow was calculated by summing the right and left pulmonary arterial flow. Systemic blood flow was calculated by adding the flow volume to the upper compartment, represented by the superior vena cava flow, to the flow volume to the lower compartment, represented by the blood flow through the DAO.

Angiography

Magnetic resonance angiography was performed with suspended ventilation if tolerated by the patient’s condition. A 3-dimensional fast spoiled gradient refocused echo sequence in the coronal orientation was used

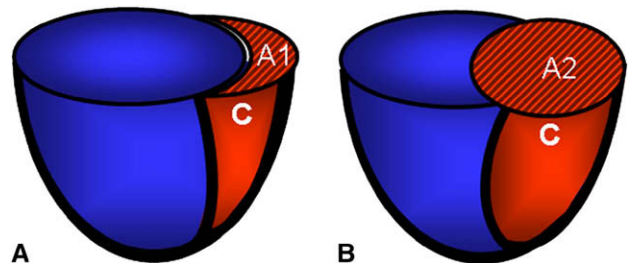


FIGURE 2. Observed (A) and potential (B) left ventricular volumes: Assuming that the left ventricle is compressible but otherwise noncompliant, the endocardial circumference (C) in any short-axis plane should remain constant, irrespective of the filling state, whereas the encircled areas (A₁ and A₂) depend on the shape of the left ventricle. The potential volume can be calculated from the area and the thickness of the imaging slices.

with the following parameters: Minimum echo and repetition times, flip angle 30°, bandwidth 31.25 kHz, 33 to 45 partitions, slice thickness 1.5 to 3.0 mm, minimum field of view (160–200 mm), matrix 256 × 128, 33 to 45 partitions. A bolus of 0.2 mmol/kg dimeglumine gadopentetate (Magnevist; Berlex Laboratoires, Quebec, Canada) was injected manually through an antecubital or hand vein, followed by a saline chaser. Centric ordered data acquisition was set to start 6 seconds after the contrast medium reached the DAO by using an automated bolus tracking technique. The diameters of the AAO, transverse aortic arch, aortic isthmus, DAO, and patent ductus arteriosus were measured on the reconstructed images using maximum intensity projections.

Echocardiography was performed on one of the following machines: Hewlett-Packard Sonos 5500 (Agilent; Andover, Mass), ATL 5000 (Advanced Technology Laboratories, Seattle, Wash), Vivid 7 (General Electric Medical Systems, Milwaukee, Wis), IE33 (Philips Medical Systems, Bothell, Wash). Measurements were partly obtained off-line and included LV end-diastolic diameter (LVEDD) by M-mode in the parasternal long-axis view, the diameters of the AV, MV, and tricuspid valve annuli in the parasternal long-axis and apical 4-chamber views, the diameters of the AAO, transverse aortic arch, aortic isthmus, DAO, and patent ductus arteriosus from suprasternal. Echocardiographic morphologic measurements were standardized to body surface area as z-scores on the basis of published normative data.⁴

LVEDV was estimated by the Simpson rule.⁵ Postoperative measurements included the diameters of the AV and MV annuli in parasternal long-axis and apical 4-chamber views, respectively, as well as the LVEDD.

Statistical Analysis

The MRI and echocardiographic data were analyzed following approval by the institution's ethics board. Continuous variables are presented as mean ± standard deviation and range. Proportions are presented as a frequency (percent). Univariable associations between continuous variables were assessed by the Spearman rho rank correlation test. Where appropriate, multivariable ordinary least square linear regression was used to assess preoperative echocardiographic predictors of the observed preoperative LVEDV by MRI. Serial echocardiographic measurements are presented using longitudinal data analysis plotting techniques. Moving average (Loess) curves of longitudinal data was done where appropriate with a smoothing span parameter (α) of 0.75 for the weighted least square method.⁶ All statistical analyses were done using the R statistical Package,⁷ using the Design⁸ and NLME⁹ libraries.

RESULTS

The patients' mean ages, weights, and body surface areas at the time of MRI were 10 days (2–37 days), 3.23 kg (1.70–4.40 kg), and 0.21 m² (0.14–0.25 m²), respectively. The mean study time was 62 minutes and there were no complications during any of the studies. The interval between the echocardiogram and the MRI ranged from 1 to 15 days (mean of 5 days). The preoperative LVEDV measured by echocardiography ranged from 6 to 25 mL/m² of body surface area by the monoplane (n = 20) and 10 to 30 mL/m² by the biplane Simpson method (n = 8). The MV and AV z-scores were -4.0 ± 2.0 (–6.8 to –0.2) and -4.6 ± 2.2 (–8.6 to –0.1), respectively. A ventricular septal defect was present in 2 of the HLHC patients.

Comparison Echocardiography—MRI

Applying the monoplane Simpson method for echocardiographic volumetry (Figure 3, A), the echocardiographic

and MRI LVEDV measurements were 16.0 ± 7.0 mL/m² and 33.5 ± 15.5 mL/m², respectively ($\rho = 0.16$, $P = .08$, n = 20). Using two echocardiographic imaging planes (Figure 3, B), the echocardiographic and MRI volumes were 21.1 ± 7.5 mL/m² and 30.6 ± 13.6 mL/m², respectively ($\rho = 0.2$, $P = .3$, n = 8).

The other anatomic measurements by echocardiography and MRI are shown in Table 1. MV annulus z-score in the echocardiographic 4-chamber view was an independent predictor of LVEDV observed by MRI ($P = .02$), whereas AV z-score and LVEDD z-score were not.

Potential LV Volumes by MRI

Every patient had a bigger potential than observed LVEDV (Figure 4). The mean potential increase of LVEDV was 8.8% for patients with AS, 35.0% for children with AVSD, and 23.0% in patients with HLHC.

Aortic Blood Flow and Shunt Volumes

PCMRI data were available in 18 of 20 patients. The measurements and calculations of the PCMRI flow analysis are given in Table 2. Blood flow volume in the ascending aorta did not correlate with LVEDV by echocardiography or MRI ($\rho = 0.01$, $P = .7$; $\rho = 0.04$, $P = .5$, respectively). All patients had antegrade flow in the AAO. In those with a subsequent BV repair, all of whom survived the early postoperative period, the AAO flow was 1.63 ± 0.57 L · min⁻¹ · m⁻² with a minimum of $1.0 \pm$ L · min⁻¹ · m⁻² in one patient. There was no correlation between the AAO flow and AV annulus z-score ($\rho = 0.003$, $P = .83$), MV annulus z-score ($\rho = 0.02$, $P = 0.69$), or AAO diameter z-score ($\rho = 0.007$, $P = .74$) by echocardiography.

Outcome

Surgical decision-making was based on multiple factors, including the patients' age and weight, their clinical status, associated cardiovascular and other lesions, imaging, as well as parental preference. In particular, in the absence of other contraindications toward a BV repair, if the MRI showed an actual LVEDV of more than the previously published cutoff of 20 mL/m², we felt encouraged to pursue a BV strategy, especially if the potential LVEDV was significantly larger. Likewise, if the MRI demonstrated that the LV outflow tract was capable of handling more than 1.0 L · min⁻¹ · m⁻² preoperatively, it seemed likely that a sufficient cardiac output would pass through it after an intervention. In the third patient of this series, hemodynamic measurements were performed intraoperatively to confirm the adequacy for BV repair.

Eighteen of 20 children underwent surgery or an intervention. One patient (HLHC, 1.70 kg, LVEDV 12.8 mL/m² by MRI) was discharged home under compassionate care by parental choice and another patient (HLHC, 2.30 kg, LVEDV by MRI 30.8 mL/m², LV and RV ejection

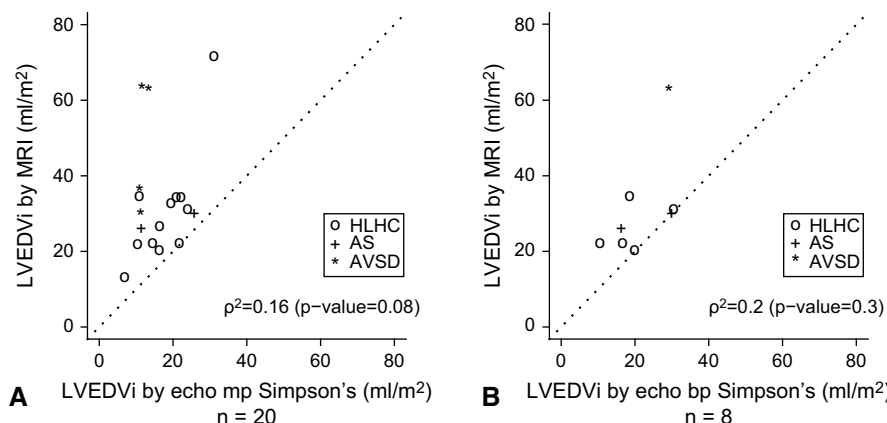


FIGURE 3. Left ventricular end-diastolic volume (*LVEDV*) measured by echocardiography versus magnetic resonance imaging (*MRI*), indexed to body surface area. **A**, Echocardiographic volume measurements, using the monoplane (*mp*) Simpson method from an apical 4-chamber view. **B**, Biplane (*bp*) echocardiographic volumetry from the apical 4-chamber and 2-chamber views. *AS*, Aortic stenosis; *AVSD*, atrioventricular septal defect; *HLHC*, hypoplastic left heart complex.

fractions 18% and 29%, respectively) died without an intervention at 6 weeks of age owing to ventricular dysfunction and periventricular leukomalacia diagnosed on day 2 after birth that precluded heart transplantation. Two patients were palliated with a UV strategy: one patient with critical *AS*, an *MRI LVEDV* of 44.1 mL/m², but decreased ejection fraction, endocardial fibroelastosis, and a small and dysplastic *MV* (z-score -2.3); the other with an unbalanced *AVSD*, an *LVEDV* of 20.7 mL/m², *LV* outflow tract obstruction, a single left papillary muscle, and decreased ventricular function with an ejection fraction of 44% (Figure 1). This patient eventually underwent a hybrid procedure (surgical pulmonary artery banding and ductal stenting in the catheter laboratory) at 3 weeks of age and a *Damus-Kaye-Stansel* anastomosis and bidirectional cavopulmonary anastomosis at 7 months. Postoperatively, central pulmonary artery obstruction led to a cardiac arrest followed by a cerebral ischemic injury and death. Two patients with critical *AS* underwent balloon valvuloplasty.

TABLE 1. Comparison of echocardiographic and magnetic resonance imaging measurements (mean ± standard deviations)

	Echocardiography	<i>MRI</i>	<i>P</i> value
<i>LVEDV</i> bp (mL/m ²)	21.1 ± 7.5	30.6 ± 13.6	0.3
<i>LVEDV</i> mp (mL/m ²)	16.0 ± 7.0	33.5 ± 15.5	0.08
<i>AV</i> annulus	4.9 ± 1.0	5.9 ± 1.0	0.006
<i>MV</i> annulus	7.5 ± 1.5	8.9 ± 1.2	0.01
<i>TV</i> annulus	12.8 ± 2.3	14.1 ± 2.3	0.14
<i>AAO</i> (mm)	6.5 ± 1.4	5.9 ± 1.2	0.4
Transverse arch (mm)	3.5 ± 0.9	3.6 ± 1.1	0.9
Isthmus (mm)	3.2 ± 1.1	3.3 ± 1.1	0.6

AAO, Ascending aorta; *AV*, aortic valve; *bp*, biplane; *LVEDV*, left ventricular end-diastolic volume; *mp*, monoplane; *MRI*, magnetic resonance imaging; *MV*, mitral valve; *TV*, tricuspid valve.

The remaining 16 patients underwent a *BV* approach. There was no early postoperative mortality. One patient died at the age of 4 months (*AVSD*, *LVEDV* 29.1 mL/m² by *MRI*) of aspiration pneumonia. Both patients with critical *AS*, who underwent *BV* repair with balloon valvuloplasty as their first intervention, survived after balloon valvuloplasty. One of them had previously undergone an arch repair. Neither required a reintervention after the balloon valvuloplasty. According to the original¹⁰ and the recently revised score¹¹ by the Congenital Heart Surgeons Society, only one of them was predicted to have a 5-year survival advantage with this strategy (5.8% and 6.7%, respectively). In the other patient, the original and the revised models estimated 2.9% and 15.6% 5-year survival benefits with a *UV* approach. Applying the *Rhodes* score, both of these patients would have been subject to a *UV* strategy.¹² They also had an aortic root size less than 3.5 mm/m² and an *MV* annulus area less than 4.75 cm²/m², both indicating a bad prognosis after *BV* repair.¹² Merely the *LV* length/ratio predicted a successful *BV* repair in both of them. An echocardiographic *LVEDV* greater than 20 mL/m², measured by biplane Simpson, was present in 1 of the 2 patients with *AS* and 3 of 7 total survivors with a *BV* repair.¹³

Two patients (one with *HLHC* and one with *AVSD*) underwent *MV* repair as a secondary procedure; the *AVSD* patient eventually required an *MV* prosthesis. One patient with *HLHC* received subaortic fibromyectomy 13 months after the initial arch reconstruction. There were no operative deaths in either the *UV* or *BV* groups. Of the 16 patients after *BV* repair, 1 died at the age of 4 months (*AVSD*, *LVEDV* 29.1 mL/m² by *MRI*) of aspiration pneumonia. There was 1 late death in the *UV* repair group following the second stage of the repair.

The postoperative growth for the *AV*, *MV*, and *LVEDd* as monitored by echocardiography is presented in Figure 5.

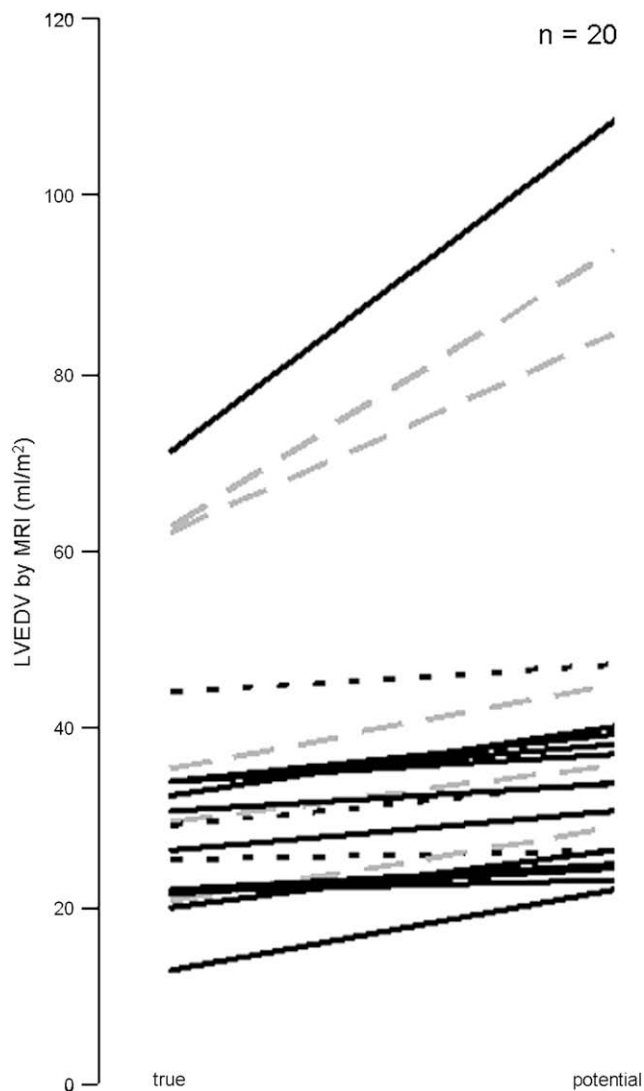


FIGURE 4. Potential left ventricular end-diastolic volumes (*LVEDV*) by magnetic resonance imaging (MRI) compared with measured true preoperative volumes. *Dotted black lines*, Patients with critical aortic stenosis; *broken gray lines*, patients with unbalanced atrioventricular septal defect; *solid black lines*, patients with hypoplastic left heart complex.

DISCUSSION

Predictors of early postoperative outcome must address whether the LV, after BV repair, is able to generate enough cardiac output to sustain the systemic circulation. Nearly all published predictors, however, are based on 2-dimensional anatomic rather than functional, indices.¹³⁻¹⁶ At our institution, there are two indications for cardiac catheterization in patients with marginally small LVs: One is to outline anatomy that cannot be detailed by echocardiography (or MRI), which was not the case in any of the patients presented here. The second is to perform an intervention, such as aortic valvuloplasty or ductal stenting during a hybrid procedure. Ideally, however, only unequivocal candidates for an inter-

TABLE 2. Flow measurements and shunt calculations

	Mean	SD
AAO flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	1.53	0.54
rSVC flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	1.16	0.56
ISVC flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	0.05	0.18
DAO flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	0.98	0.42
RPA flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	3.07	1.40
LPA flow ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	2.14	1.12
Q_p (RPA flow + LPA flow) ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	5.21	2.44
Q_s (SVC flow + DAO flow) ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	2.18	0.83
Q_p/Q_s	2.86	1.81
Total left-to-right shunt ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)*	3.04	2.78
PDA flow (left to right) ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)	-0.46	0.95
Intracardiac left-to-right shunt ($L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$)†	3.51	2.39

AAO, Ascending aorta; DAO, descending aorta; LPA, left pulmonary artery; RPA, right pulmonary artery; PDA, persistent ductus arteriosus; Q_p , pulmonary blood flow; Q_s , systemic blood flow; r/SVC, right/left superior vena cava. *Calculated using $Q_p - Q_s$. †Calculated as $Q_p - Q_s - \text{PDA flow (left to right)}$.

vention should undergo cardiac catheterization. The difficulty in establishing a uniformly recognized predictor of outcome after surgery, along with the observation that it is not always the patients with the smallest LV structures that have an unsuccessful BV repair, suggest that a different approach is needed.² In this study, we attempted to overcome these limitations and compress the size of the marginal decision-making zone within the spectrum of borderline hypoplasia of the LV by using 3-dimensional anatomic and functional parameters of native LV performance. This study is the first to show that functional MRI in these neonatal patients is feasible and may be useful.

LV Output as the Result of Size and Function

Cardiac output is the result of a complex interplay of heart rate, LV preload, ventricular interaction, myocardial contractility, and systemic resistance. Notwithstanding the ductal contribution to the systemic circulation, the presence of forward flow in the ascending aorta, seen echocardiographically, is reassuring when BV repair is considered.¹⁷ Our group has recently shown that a systemic flow of only $1.8 \pm 0.6 L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$ was present in survivors early after the Norwood operation.¹⁸ The survivor with a BV repair in our cohort demonstrated preoperatively that a flow comparable with that number ($1.63 \pm 0.57 L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$) could pass through the LV outflow tract and AAO, with a minimum flow as low as $1.0 L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$. Therefore, although our data cannot provide a threshold below which preinterventional aortic flow may be inadequate, we chose $1.0 L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$ pragmatically in this series.

PCMRI is the method of choice for flow volume measurements.^{19,20} If the region of interest contains at least 16 pixels, the inaccuracy is less than 10%.^{21,22} In our study, the smallest aortic cross-sectional area was 13 mm^2 , seen in the smallest neonate, weighing 1.70 kg. In this child, the region of

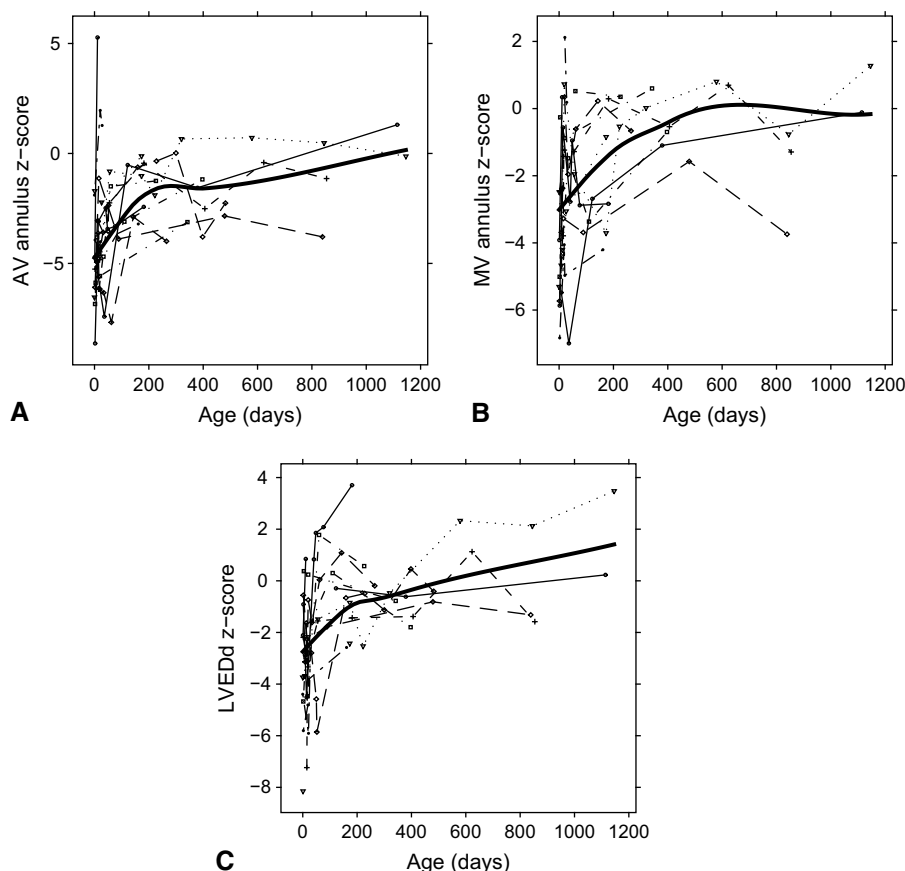


FIGURE 5. Postoperative growth after biventricular repair, monitored by serial echocardiograms: The first data point is the preoperative size; all subsequent points are postoperative measurements. Z-scores for (A) aortic valve annulus, (B) mitral valve annulus (excluding patients with atrioventricular septal defect), and (C) left ventricular end-diastolic diameter.

interest contained 28 pixels at a spatial resolution of 1.00×0.47 mm. This report is the first of noninvasive shunt calculations in neonates.^{23,24} We recently assessed the accuracy of PCMRI in normal adult volunteers, using the same magnet as for this study, and found an error of only 0.02 ± 0.2 $L \cdot \text{min}^{-1} \cdot \text{m}^{-2}$ when comparing the measured flow through the DAO with the measured flow (AAO—superior vena cava flow). This deviation was equivalent to $0.69\% \pm 5.96\%$ of systemic flow (H. W. Goo, MD, unpublished data, 2007). In the present patient cohort, characterized by a labile balance of the pulmonary and systemic blood flows, it is important to obtain flow data from various locations as quickly as possible and to keep the conditions for continuous breathing or ventilation and the patient's fluid status as constant as possible.

Echocardiography Underestimates LV Volume

In older patients, MRI is the established gold standard for ventricular volumetry inasmuch as it does not rely on a presumed shape of the ventricle, in contrast to the echocardiographic Simpson method that assumes a bullet-shaped LV, with a circular cross section.^{5,25-29} The fact that the shape

of the LV in patients with a borderline LV is abnormal is almost certainly the reason for the inaccuracy of this technique, when compared with MRI (Figure 3).³⁰ This error leads to an underestimation of the true volume if the septum is bowing toward the left side, which is amplified when the LV diameter is measured only in the 4-chamber plane instead of a biplane assessment (Figure 3). None of the patients received an MRI and an echocardiogram on the same day. In some of them, the two tests were 2 weeks apart. This, through ventricular growth and altered shunts and loading conditions, might have contributed to the discrepancy between echocardiography and MRI-derived volumetrics. MV annulus size predicts survival in patients with left-sided obstructive lesions.^{12,31,32} In a report by Schwartz, Gauvreau, Geva,³¹ echocardiographic MV annulus diameter and LVEDV correlated in children with multiple left-sided obstructive lesions ($\rho = 0.47$, $P < .001$). Our data confirm that echocardiographic measurements of LV inflow, but not outflow size or end-diastolic dimension, predict true LVEDV as measured by MRI.

MRI measurements of the AV and MV were significantly larger than echocardiographic measurements (Table 1).

Echocardiography, especially in neonates who typically have very good ultrasound windows, appears superior to MRI for intracardiac structures, owing to better temporal resolution and tissue/blood contrast in echocardiography versus white blood MRI.

MRI Changes Surgical Decision-making

Conversion to a UV circulation after a failed BV repair carries a high mortality, whereas the opposite sequence of interventions appears to have a better result.^{11,12,33} A UV circulation, on the other hand, may limit the child's functional outcome. The hitherto largest study by the Congenital Heart Surgeons Society estimated that half of all patients with critical AS who underwent a primary BV repair should have had a Norwood procedure, whereas 20% of those who did have a UV repair would probably have survived after BV repair.¹⁰ The Society's original score to predict 5-year survival benefits of UV over BV repair was recently revised.¹¹ In both the original and the new version, 1 of 2 patients with AS and a successful BV repair in this cohort would not have been given that chance.

Concluding that the adverse effects of inflow and outflow hypoplasia and a small LV are cumulative, Rhodes and colleagues¹² combined the diameters of the aortic root and MV annulus and the LV length in a score to predict survival after BV repair in patients with AS. In our cohort, it would have mandated a Norwood operation in both patients with AS who underwent successful BV correction.

The Rhodes score, however, is not applicable to patients with HLHC or unbalanced AVSD.³⁴ In these patient groups, discriminating parameters are scarce, and although several reports of survivors of BV repairs with smaller ventricles undermine this cutoff, an LVEDV of at least 20 mL/m², measured by echocardiography or x-ray angiography, continues to be commonly used as a prerequisite for BV repair.^{31,35} Interestingly, these studies used echocardiography for their volumetry, whereas the threshold of 20 mL/m² was originally established angiographically.³⁶ In our group of patients with BV repair, none of whom died perioperatively, the smallest LVEDV was 20 mL/m² by MRI. More than half of them would have undergone UV repair, applying this still widely used criterion to their echocardiographic measurements. Thus our data suggest that an LVEDV of 20 mL/m² or more is sufficient to undergo BV repair whereas echocardiographically obtained volumes are less meaningful and may be considerably smaller.

In patients with unbalanced AVSD or HLHC, who have RV pressure and/or volume overload, a mere expansion of the LV can lead to a substantial increase in volume, as expressed by a large difference between the true and the potential LVEDV (Figures 2 and 4).^{3,37} In patients with an actual LVEDV of 20 mL/m² or less but otherwise encouraging anatomy and function, a large potential increase in ventricular size after optimized filling conditions is reassuring

toward a BV repair. In contrast, in critical AS, which is characterized by a markedly increased afterload, the LV retains a near ideal shape. Therefore, the potential volume increase in these patients is small, even if RV and LV loading conditions are optimized. Expansion and an altered shape of the LV immediately postoperatively, as well as continued growth, presumably contribute to the increase in size of LV structures after BV repair (Figure 5).

Of note, 2 of our patients underwent UV repair despite an LVEDV greater than 20 mL/m², because valvular anatomy and/or myocardial function at the time of surgery was deemed inadequate for a BV approach, underscoring the continued importance of associated lesions, especially of the LV inflow. Han and colleagues³⁸ recently showed that after neonatal intervention for critical AV stenosis the potential of the MV annulus for catch-up growth is smaller than that of the AV and LV diameter.

It is important to appreciate that long-term survival does not necessarily exclusively reflect appropriate selection, as continuing growth and physiologic remodeling are necessary in these BV repairs. Our early data and those of others regarding expansion and growth of LV structures are reassuring in this regard, but careful follow-up will be required.³⁸

The ideal outcome for any patient is a serial circulation using two ventricles, free of repeat interventions and late complications. The recent analysis of 362 patients with critical AS by the Congenital Heart Surgeons Society suggests that 56% of patients who are submitted to a BV repair would have had a long-term survival advantage from a UV approach, and that erring on the side of a BV repair has disastrous effects in a large proportion of these patients.¹¹

LIMITATIONS

The principal limitation of this study is the relatively small number of patients, making it difficult to test for statistical significance. Therefore, a lack of an association in our analysis does not exclude it. Cardiac MRI is now part of the standard of care in patients with borderline small LVs in whom there is doubt whether to attempt BV or UV at our institution. The present study contains a selection bias inasmuch as not all patients with comparable pathologic anatomy underwent an MRI in the early phase of this study.

This study was not designed to look at outcome for particular lesions or interventions. The patients were selected on the basis of having undergone a preoperative MRI to assess LV size. There were no perioperative deaths after the initial BV repair and no patient had to be converted into a UV circulation. Therefore, MRI-derived risk factors for unsuccessful BV repair could not be isolated from our data. Prospective studies of a greater cohort with more adverse outcomes after BV repair are needed to answer this question.

A biplane echocardiographic volume assessment in all patients would have been desirable as it is the most common method and more accurate than a monoplane assessment.

Early postoperative MRI examinations are desirable to detail how much of the potential volume is actually recruited by loading the LV and unloading the RV.

In conclusion, MRI volumetric and flow measurements in neonates with borderline hypoplasia of the LV are feasible. Echocardiographic volumetry underestimates LV volume, and indices based on anatomy alone may not adequately predict survival after BV repair. Consequently, assessment of the LV volume and cardiac output by MRI may change surgical decision-making regarding UV versus BV repair.

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