

The Functional Intraoperative Pulmonary Blood Flow Study Is a More Sensitive Predictor Than Preoperative Anatomy for Right Ventricular Pressure and Physiologic Tolerance of Ventricular Septal Defect Closure After Complete Unifocalization in Patients With Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collaterals

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Background—The objective was to determine if intraoperative pulmonary artery (PA) flow studies after complete unifocalization correlate with postrepair hemodynamics for pulmonary atresia (PA), ventricular septal defects (VSD), and major aortopulmonary collaterals.

Methods and Results—Twenty patients (median age, 8 months; weight, 7.9 kg) underwent unifocalization between 2003 and 2008. A functional PA flow study was achieved by cannulating the unifocalized central PA before intracardiac repair and increasing flow incrementally to 2.5 L/min per m². Mean PA pressure (mPAP) was measured. The intent was to close the VSD for a mPAP of <30 mm Hg. Right ventricular systolic pressure (RVSP) and systemic systolic pressure were recorded. Total incorporated pulmonary segments, pulmonary segment artery ratio (ratio of incorporated segments to 18), and total neopulmonary artery index (the sum of major aortopulmonary collaterals and native PA index) were calculated. The VSD was successfully closed in 18 patients (90%). One attempted closure required an intraoperative fenestration. The study mPAP correlated with RVSP ($\rho=0.72$; $P=0.0027$) and RVSP/systemic systolic pressure ($\rho=0.67$; $P=0.0063$). Total neopulmonary artery index had a nonsignificant negative correlation with RVSP ($\rho=-0.42$; $P=0.079$). Total incorporated pulmonary segments and pulmonary segment artery ratio were not correlated. Flow study mPAP had the highest accuracy in predicting successful VSD closure: area under the receiver-operator curve (0.83) versus total neopulmonary artery index (0.42), pulmonary segments (0.35), and pulmonary segment artery ratio (0.33).

Conclusions—The intraoperative pulmonary flow study predicted postoperative physiology significantly better than did standard anatomic measures. Conventional measures should be used with caution when determining the possibility for complete repair. (*Circulation*. 2009;120[suppl 1]:S46–S52.)

Key Words: aortopulmonary collaterals ■ blood flow ■ heart defects, congenital ■ perfusion ■ pulmonary atresia

Pulmonary atresia (PA) with a ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCA's) is a congenital heart malformation with varying iterations of segmental pulmonary blood supply.^{1,2} Surgical management goals have been to achieve a central mediastinal confluence for all sources of pulmonary blood flow (this is termed unifocalization) and then to complete the repair with closure of the VSD and placement of a right ventricle-to-pulmonary artery connection. Surgical repair can be performed in 1 stage or in multiple stages of unifocalization with

subsequent intracardiac repair.^{3,4} The traditional staged approach has the disadvantage of leaving MAPCA's and their lung segments in their native state for months to years. There can be development of MAPCA stenosis or even potential injury to the pulmonary vascular bed caused by unrestrictive systemic pressure and blood flow if MAPCA stenosis does not occur. In 1995, Hanley et al⁵ proposed 1-stage complete unifocalization and VSD closure aiming to incorporate all segments of blood supply before stenosis developed in the MAPCA's and before pulmonary vascular obstructive disease

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Presented in part at American Heart Association Scientific Sessions 2008, November 8–12, 2008, New Orleans, La.

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Circulation is available at <http://circ.ahajournals.org>

DOI: 10.1161/CIRCULATIONAHA.108.844084

occurred. This approach had been applied for >90% of the patients with PA/VSD/MAPCA's in whom two-thirds of the patients actually had VSD closure.⁶ Both the single-stage and multiple-stage approaches have yielded varying and imperfect outcomes for complete repair: multiple-staged complete repair, 29% to 61%; single-stage complete repair, 50% to 85%.^{3,4,6,7}

Postoperative right ventricular systolic pressure (RVSP) is a marker for whether VSD closure can be physiologically tolerated. If the VSD is closed and there is subsequent suprasystemic RV pressure, morbidity and mortality are high.⁷⁻⁹ Preoperative anatomic evaluation has been used to determine which patients will have a successful complete repair. Data utilized have been the total number of incorporated pulmonary segments or a pulmonary segment artery ratio (PSAR) after unifocalization^{4,10,11} More recently, a combined area index for all MAPCA's and the central PA, ie, total neopulmonary artery index (TNP AI), has been suggested.⁹ Preoperative anatomic findings have not always predicted postoperative functionality.

A functional measurement of pulmonary vascular performance may be more predictive of post reconstruction pressures. Reddy et al⁹ described an intraoperative pulmonary blood flow study that measured mean PA pressure simultaneous to pumping blood flow through the unifocalized pulmonary arteries at a cardiac index of 2.5 L/min per m². A mean PA pressure <30 mm Hg was thought to allow successful VSD closure at a reasonable RV pressure.

We adopted the functional intraoperative pulmonary blood flow study to determine the possibility of VSD closure at complete unifocalization. We hypothesized that the mean PA pressure obtained from the functional intraoperative pulmonary flow study would better-correlate with postoperative RSVP as compared to anatomic measurements. That is to say, a functional measurement would better-measure functionality than imputing functionality from anatomy. As such, all patients underwent a strategy for complete unifocalization with or without a subsequent complete repair regardless of presenting anatomy. The hypothesis is evaluated in the following study.

Materials and Methods

We reviewed all patients with PA/VSD/MAPCA's who underwent a cardiac surgical procedure (1-stage or multiple-stage unifocalization and VSD closure) between January 2003 and August 2008. The Research Ethics Board at the Hospital for Sick Children approved the study and waived the requirement for patient consent. Twenty patients were identified. All patients were included in the study. All patients underwent a functional intraoperative pulmonary blood flow study and complete unifocalization, with or without VSD closure. Patients' preoperative characteristics are shown in Table 1. Of note, 1 patient had required extracorporeal membrane oxygenation after a preoperative cardiac diagnostic catheterization.

Surgical Strategy and Technique

Our surgical approach is to attempt 1-stage unifocalization and VSD closure in all patients at 4 to 8 months of age. A thoracotomy-based staged unifocalization is used if a patient has significant distal stenoses in the MAPCA's that cannot be accessed via a sternotomy. Diminutive central PA's (<2 mm) are rehabilitated with an autologous main PA-to-aorta central shunt (Mac Shunt).³

One-stage unifocalization and repair was performed through median sternotomy as described by Hanley et al.^{5,10} The central PA's were dissected and mobilized. The posterior mediastinum was

Table 1. Patients Characteristics

Patients	20
Male/female	12/8
Median age at repair, mon	7.8 (range 2–197)
Median body wt at repair, kg	7.9 (range 2.7–64)
DiGeorge syndrome	3 (15%)
Microdeletion of chromosome 22q11	1 (5%)
Native pulmonary artery	
Confluent	13 (65%)
Well-developed	7 (35%)
Diminutive or small	6 (30%)
Nonconfluent	4 (20%)
Absent	3 (15%)
Median N of MAPCA's	3 (range 1–4)
Staged palliation	6 (30%)
Mee shunt	2
Unilateral unifocalization and shunt	2
Modified Blalock-Taussig shunt	1
Mee shunt followed by unilateral unifocalization	1

dissected creating a window to dissect the MAPCA's. After controlling all MAPCA's, cardiopulmonary bypass (CPB) was initiated and the proximal portions of the MAPCA's were snared. The central PA's were opened from hilum to hilum. Each MAPCA was divided adjacent to the aorta and shortened to the appropriate length, excluding any stenotic area. Each MAPCA was then anastomosed to the side or back wall of the central PA (if present) using 7-0 or 8-0 polypropylene sutures (Ethicon, Inc, Somerville, NJ). After completing unifocalization, the anterior/caudad aspect of the central PA's were augmented from hilum to hilum with an autologous pericardial patch. Even if the mediastinal PA's were of reasonable size, the pericardial patch was added. The distal conduit was then sewn into the opened patch so that there was no distortion or stenosis created in the mediastinal PA's from the conduit anastomosis. We evolved to use glutaraldehyde-treated autologous pericardium for the central PA patch. In cases of nonconfluent central PA's, a tissue-to-tissue anastomosis was achieved in the posterior mediastinum.

Functional Intraoperative Flow Study and VSD Decision

After completing unifocalization and central PA patch arterioplasty, an intraoperative functional pulmonary blood flow study was achieved by inserting an additional arterial cannula in the reconstructed central PA's (Figure 1). The flow study was performed simultaneous to systemic total CPB and cardioplegic arrest. Complete decompression of the left atrium, with multiple pump suction devices, and normal ventilation was achieved. The flow study was initiated at ≈20% of the final planned 2.5 L/min per m². A 20-G needle, attached to a pressure line and transducer, was inserted into the reconstructed central PA for continuous mean PA pressure measurement. Flow was incrementally increased until the final indexed rate was achieved. The intent was to close the VSD if the mean PA pressure, at an index of 2.5L/min per m², was ≤30 mm Hg. An RV-to-PA continuity was established with either a pulmonary homograft or a bovine jugular vein conduit (Contegra pulmonary valved conduit; Medtronic, Inc, Minneapolis, Minn). An intraatrial fenestration of 3 to 4 mm was left for all but those having very low PA pressures with the flow study.

RVSP was measured after separation from CPB and completion of modified ultrafiltration. A 20-G needle attached to monitoring equipment was inserted into the right ventricle through the anterior wall for pressure tracing and measurement. Systemic systolic blood

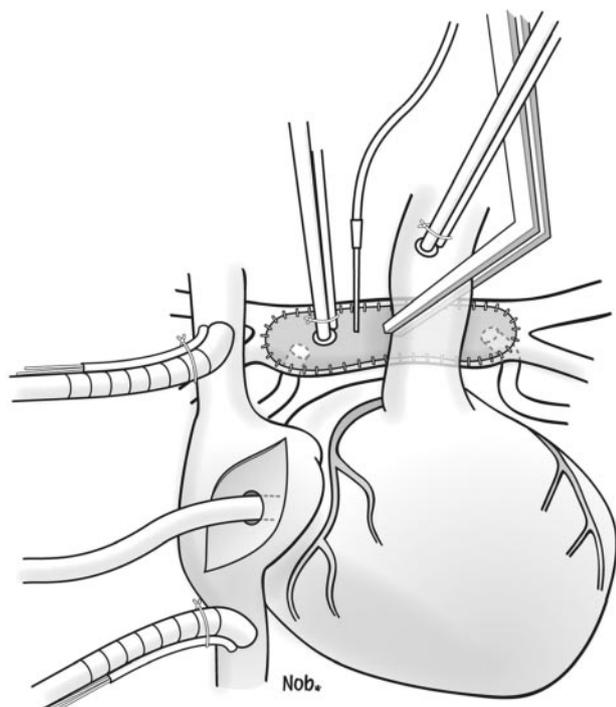


Figure 1. Intraoperative functional pulmonary flow study. An additional cannula is inserted into the completely unifocalized and reconstructed central PA. The pulmonary vasculature is perfused with an incrementally increasing flow to a rate of 2.5L/m² per minute under cardioplegic cardiac arrest. The left atrium is fully vented and the lungs are adequately ventilated.

pressure (SBP) obtained from a radial or femoral artery was simultaneously recorded. Direct needle-based aortic pressure was taken if needed for clarity of accuracy. A sequential pressure measurement in the central pulmonary artery adjacent to the attachment of the RV-PA conduit was also taken. The RVSP-to-SBP ratio was calculated.

Analysis of Anatomic Predictors

The preoperative angiograms of all patients were reviewed. Measurements were performed by an experienced pediatric cardiac radiologist who was blinded to the clinical profiles of the patients (C.M.). Three anatomic observations/calculations were achieved. Total incorporated pulmonary segments were calculated as the sum of the number of pulmonary segments supplied by the MAPCA's that had eventually been unifocalized and the pulmonary segment supplied by the native central PA's. Pulmonary segment artery ratio was calculated as the counted total incorporated pulmonary segments divided by total pulmonary segments of the patient, which are 18 in most of the patients. Total neopulmonary arterial index was calculated as the sum of the total MAPCA index and the native pulmonary artery index.⁹ The sizes of the central PA were measured just before their bifurcation, as described by Nakata et al.¹² The sizes of MAPCA's were

measured immediately distal to the likely site for surgical unifocalization and beyond any central stenosis if present. Indices were determined by calculating cross-sectional area divided by body surface area.

Statistical Analysis

Data are presented as means±SD. The level of statistical significance was set at $P=0.05$. Correlations between preoperative and postoperative variables were analyzed by Spearman rank correlation. The accuracy of the anatomic and functional indicators in predicting the possibility of VSD closure was analyzed by area under the receiver operator characteristics curve. Freedom from death and from reintervention were analyzed using Kaplan-Meier survival analysis. Differences between early and late postoperative RVSP were analyzed by Student t test.

Results

Six (30%) out of 20 patients received staged palliation for either rehabilitation of the diminutive central PA ($n=4$; 20%) or distal MAPCA stenosis ($n=2$; 10%). Therefore, 14 (70%) out of 20 patients had 1-stage unifocalization. For the studied procedure, aortic cross-clamp time was 82 ± 44 minutes (median, 68; range, 27–224). Cardiopulmonary bypass time was 208 ± 86 minutes (median, 209; range, 67–386). RV-to-PA continuity was established with a bovine jugular vein conduit in 14 (70%) patients, a homograft was used in 5 (25%) patients, and a transannular patch using autologous pericardium was used in 1 (5%) patient.

Total incorporated pulmonary segments were 16.1 ± 1.9 (median, 16; range, 12–18). Pulmonary segmental artery ratio to 18 was 0.89 ± 0.11 (median, 0.91; range, 0.66–1). Total neopulmonary artery index was 141 ± 68 mm²/m² (median, 120; range, 62–333). Six (20%) patients had TNPAI <100 mm²/m².

The mean PA pressure obtained from the functional intraoperative pulmonary blood flow study was 21.8 ± 6.2 mm Hg (median, 23; range, 11–31). Complete closure of the VSD was performed in 19 (95%) patients. One (1/19) underwent fenestration in the operating room for suprasystemic RV pressures (final total VSD closed=18/20, 90%). Three (15%) had a mean PA pressure of ≥ 30 mm Hg by the flow study. Of those, 1 had a mildly restrictive fenestrated VSD patch placed (mean PA pressure on flow study=30 mm Hg and 16 incorporated segments), and 2 had VSD closure (mean PA pressure of 30 and 31 mm Hg by flow study and 18 incorporated segments; Table 2). The latter 2 had RVSP/SBP ratios of 0.78 and 0.97, respectively. The decision to close those VSD was based on the marginal nature of the flow study combined with the fact that their anatomy was thought to be favorable. The respective last follow-up RVSP/SBP ratios are 0.51 and 0.32, respectively. The 1 patient who had a second cardiopulmonary bypass run for VSD fenestration initially had a

Table 2. Characteristics of the Patients Who Had Mean PA Pressure >30 mm Hg or VSD Fenestration

Patient	Total Incorporated Segments	PSAR	TNPAI	mPAP at Flow Study	RVSP	RVSP/SBP Ratio	VSD	RVSP/SBP at Follow-Up
1	16	0.89	184.5	25	85	1.3	Closed then fenestrated after having suprasystemic RVSP	0.94
2	18	1	103.1	31	73	0.97	Closed	0.32*
9	18	1	76.2	30	55	0.78	Closed	0.51
12	15	0.83	62.8	30	NA	NA	Electively fenestrated	NA

NA indicates not available

*After catheter intervention.

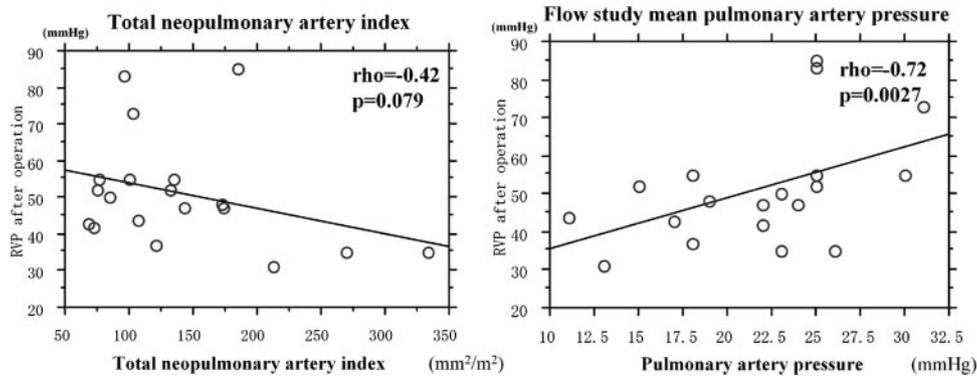


Figure 2. Correlation between TNPAI or the mean PA pressure on the flow study and RVSP after VSD closure. There was a nonsignificant trend toward negative correlation between TNPAI and RVSP. There was a strong positive correlation between the mean PA pressure, on the flow study, and RVSP.

flow study mean PAP of 25 mm Hg. The total incorporated segments in that patient were 15. The RVSP after the initial separation from bypass was 85 mm Hg, with the RVSP/SBP being 1.3, thus making the decision for fenestration clear.

An intraatrial fenestration was left in 15 (75%) of the patients. The mean RVSP after VSD closure was 51 ± 15 mm Hg (median, 48; range, 31–85), and RVSP/SBP ratio was 0.63 ± 0.21 (median, 0.58; range, 0.41–1.3).

Correlation Between Anatomic Predictors and PA Pressure on the Flow Study

There was no correlation between total incorporated pulmonary segments ($\rho=0.246$; $P=0.29$) or PSAR ($\rho=0.25$; $P=0.31$) and the mean PA pressure by the flow study. There was a weak negative correlation between TNPAI and the flow study PA pressure, ie, the higher the TNPAI, the lower the mean PA pressure ($\rho=-0.324$; $P=0.16$).

Correlations Between Anatomic or Functional Predictors and Postoperative RV Pressure

There was a positive correlation between the mean PA pressure on the flow study and the postoperative RVSP ($\rho=0.72$; $P=0.0027$; Figure 2). There was a positive correlation between the mean PA pressure on the flow study and RVSP/SBP ratio ($\rho=0.67$; $P=0.0063$; Figure 3). Total incorporated pulmonary segment and PSAR were not correlated with the postoperative RVSP ($P=NS$). A nonsignificant trend

toward negative correlation between TNPAI and the mean PA pressure by the flow study was noted ($\rho=-0.42$; $P=0.079$). No correlation between total incorporated pulmonary segments, PSAR, or TNPAI and RVSP/SBP ratio ($P=0.37$) was seen.

The area under the receiver operator characteristics curve was highest for correlation to the mean PA pressure by the flow study (0.83), followed by TNPAI (0.42), then total incorporated pulmonary segments (0.35), and, last, PSAR (0.33).

Clinical Outcomes and Follow-Up

There was no early death. No patient required early reoperation. Four patients underwent delayed sternal closure. One patient had an episode of cardiac arrest within hours of arrival in the intensive care unit. Standard resuscitation including chest opening and delayed sternal closure were successful. Median duration of intensive care unit stay and hospitalization after unifocalization and VSD closure were 5.5 days (mean, 12; range, 2–105) and 10 days (mean, 28; range, 4–183), respectively. There was 1 (5%) late death. The patient had a history of arrhythmia related to poor preoperative ventricular function. One-stage unifocalization and complete repair were performed subsequent to heart failure treatment with a β -blocker. Postoperative RVSP/SBP ratio was 0.44. Subsequently, PA stenosis developed. Hemodynamic decompensation followed a catheter-based intervention, which was also associated with poor ventricular function. Actuarial survival at 1 year is 94% (Figure 4A).

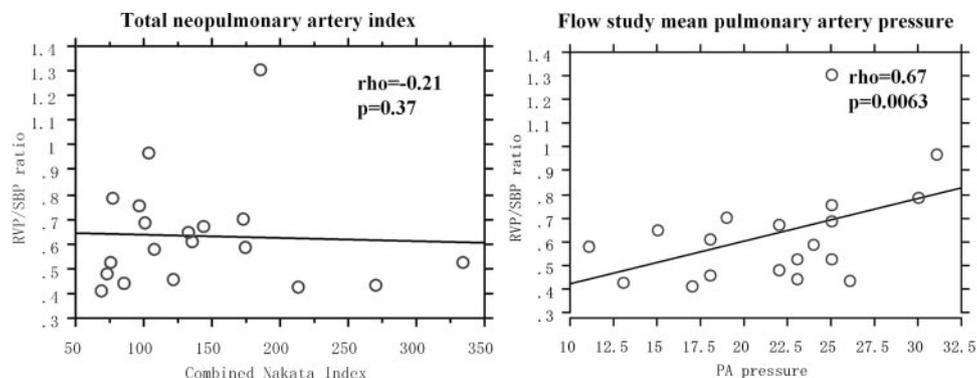


Figure 3. Correlation between TNPAI or the mean PA pressure on the flow study and RVSP/SBP ratio after VSD closure. There was a positive correlation between the mean PA pressure on the flow study and RVSP/SBP, whereas no correlation was noted between TNPAI and the mean PA pressure on the flow study.

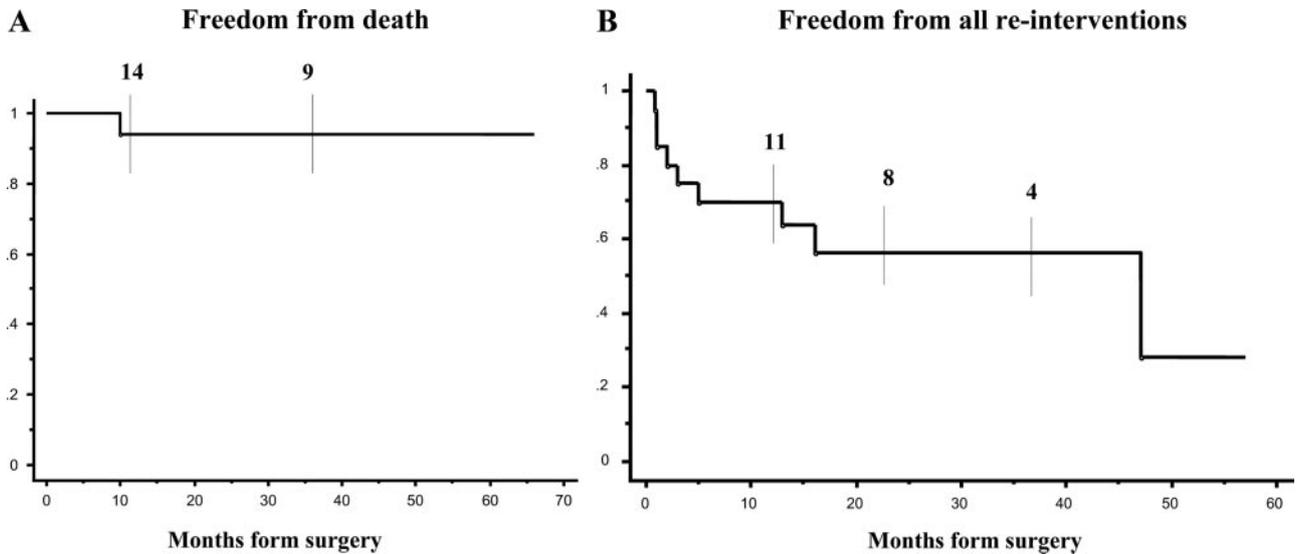


Figure 4. Kaplan–Meier survival curves showing the freedom from death (A) and freedom from all reinterventions. Actuarial survival rate at 1 year after complete unifocalization was 94%. Actuarial freedom from all reintervention rate was 70% at 1 year and 57% at 3 years.

The median follow-up period was 31 months (mean, 30; range, 8–66). Ten (50%) underwent 11 surgical or catheter-based reinterventions as follows: balloon angioplasty for branch PA stenosis (n=8), coil embolization for small collaterals causing hemoptysis (n=1), and suspension of the PA or aorta for bronchial stenosis (n=2). Actuarial freedom from all reintervention was 70% at 1 year and 57% at 3 years, respectively (Figure 4B). Preinterventional and postinterventional RVSP/SBP ratio was $79.5 \pm 14.9\%$ and $67.3 \pm 18.0\%$, respectively (n=4).

RVSP and RVSP/SBP ratio estimated by echocardiography at discharge and at the latest follow-up were obtained in 15 (79%) patients. Five patients did not have tricuspid regurgitation; therefore, quantitative estimation of RVSP was not possible. A positive correlation between the mean PA pressure by flow study and the estimated RVSP at discharge ($\rho=0.540$; $P=0.038$), and RVSP/SBP ratio ($\rho=0.524$; $P=0.045$) was present. At the latest echocardiographic follow-up study (median, 20 months; range, 4–61 months), there was no correlation

between mean PA pressure by flow study and RVSP ($\rho=-0.08$; $P=0.77$) or RVSP/SBP ratio ($\rho=-0.11$; $P=0.70$). There was no correlation between total incorporated segments, PSAR, or TNPAI and RVSP, or RVSP/SBP at discharge ($P=NS$).

RVSP and RVSP/SBP ratio at the latest follow-up was lower than those at discharge (RVSP at discharge 54 ± 22 mm Hg versus RVSP at follow-up 46 ± 18 mm Hg; $P=0.055$; RVSP/SBP ratio at discharge, 0.64 ± 0.27 versus RVSP at follow-up 0.53 ± 0.22 ; $P=0.039$; Figure 5A,B).

Discussion

An intraoperative pulmonary blood flow study is an opportunity to obtain data on the functionality of the entire pulmonary vasculature once it has been unifocalized but before closing the VSD. It offers the possibility for more refined decision-making regarding the appropriateness of VSD closure. An incorrect decision regarding treatment of the VSD has untoward consequences. In the most severe

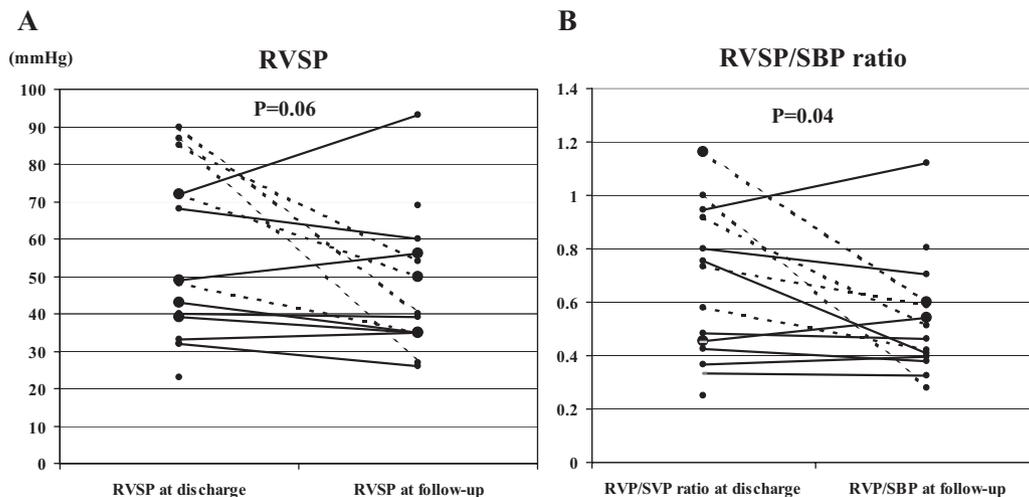


Figure 5. Changes in RVSP and RVSP/SBP ratio during follow-up. Solid lines represent the patients who did not undergo any reintervention. Dotted lines represent the patients who underwent reintervention for stenotic branch PA. RVSP and RVSP/SBP ratio were significantly lower compared to the values at discharge. RVSP was estimated by tricuspid regurgitation gradient on echocardiography.

cases of elevated pulmonary vascular resistance, separation from CPB may not be possible because of RV failure or inadequate left ventricular filling. Reddy et al.⁹ reported that VSD closures not physiologically tolerated were associated with significant acute morbidities, including severe hemodynamic instability and multiorgan injury. Failure to close the VSD when the PVR is sufficiently low can result in too much pulmonary blood flow that may require early reintervention for VSD closure or banding of the RV-PA conduit.⁹ In our patient population, the flow study mean PA pressure was highly correlated with postoperative RVSP. It thus allowed for reasonable prediction of postoperative physiology and, therefore, tolerance of the chosen end physiological state. In the present cohort, 1 of 25 patients (5%) had a VSD status when leaving the operating room which was different than what was planned based on the flow study. No patient had to subsequently return to the operating room for fenestration or VSD closure.

There are several factors influencing the mean PA pressure with a functional intraoperative pulmonary blood flow study. Segmental atelectasis associated with discontinuation of mechanical ventilation during CPB may affect pulmonary vascular resistance. The lungs should be fully recruited and adequately ventilated. All fluid/blood collected in the pleural space should be evacuated to allow full lung expansion. The left atrium should be decompressed with multiple suction catheters from the bypass machine. Our experience showed that incomplete decompression of the left atrium, such as performance of the flow study on the beating heart, resulted in significantly higher mean PA pressure than in a fully arrested and decompressed cardiac state. Also, significant leakage from the reconstructed central PA suture line needs to be repaired to allow accurate measurement of the mean PA pressure. Inadequately addressing these issues can lead to a misleading outcome for the flow study and potentially an incorrect decision regarding VSD closure. If an incorrect decision regarding VSD closure can be identified while still in the operating room, addressing the VSD appropriately at that time appears to be reasonably tolerated.

Anatomic Predictors of Tolerance to VSD Closure

In the past, TNPAI has been shown to be the best anatomic indicator of postoperative RVSP and successful VSD closure at a physiologically tolerated RVSP. Reddy et al.⁹ showed that a higher TNPAI was correlated with a more favorable postoperative RVSP/left ventricular pressure ratio. All patients who needed VSD fenestration had a TNPAI <200 mm²/m². In the present study, TNPAI had nonstatistically significant correlation with the postoperative RVSP. There was no correlation between TNPAI and RVSP/SBP ratio. The accuracy of predicting VSD closure was poor, ie, 42% (area under the receiver operator characteristics curve, 0.42).

The limitation of anatomic predictors is that anatomy does not necessarily correlate uniformly with function. If a large unobstructed MAPCA perfuses a unilateral whole-lung field, the resistance of the lung field may be normal or high, depending on how long the lung field has been exposed to the high pressure or how quickly the lung vasculature reacts to the high flow and pressure of the nonstenosed MAPCA. Nevertheless, TNPAI is

an important piece of anatomic information that has some correlation to clinical outcomes. It is also readily attainable.

Investigators from the Mayo Clinic have suggested that having <14 incorporated segments has a significant impact on total resistance and pressure of the pulmonary vasculature after unifocalization, thereby influencing the possibility of VSD closure.⁴ Total incorporated pulmonary segments did not correlate with postoperative RVSP and RVSP/SBP ratio in our study. A possible reason for this negative result is that only 4 patients had <14 unifocalized pulmonary segments. Nevertheless, successful closure of the VSD with <14 incorporated segments was achieved, indicating that that anatomic finding is not completely predictive.

Decision-Making in the Marginal Pressure Patient

Reddy et al.⁹ suggested that the VSD should be fenestrated or left open if the mean PA pressure was >30 mm Hg on the flow study. If we examine data from the present cohort, useful information can be added. In the present study there were 4 patients who either had mean pressure on the flow study >30 mm Hg or required fenestration because of suprasystemic RVSP subsequent to VSD closure (Table 2). Patient 1 had a mean PA pressure of 25 mm Hg and subsequently required VSD fenestration because of suprasystemic RVSP. The TNPAI was 184 mm²/m². The finding was not expected. The patient was the only one with a false-negative result in the study. We performed VSD closure in patients 2 and 9 despite a mean flow study PA pressure of 30 and 31 mm Hg. These 2 studies could be termed false-positives because each patient had mid-term follow-up RVSP/SBP ratio $<50\%$. The decision to close the VSD was made in conjunction with the fact that there was good central PA reconstruction, incorporation of all lung segments, and absence of severe arborization abnormalities or distal segmental pulmonary hypertension. The VSD was electively fenestrated in patient 12, who had a mean flow study PA pressure of 30. The child was 20 months of age and had absent central PA with 2 MAPCA's. One of those MAPCA's had no stenosis and fed the entire left lung. The preoperative mean MAPCA pressure was 73, indicating that there might be pulmonary vascular disease. A summarization of the decision-making strategy utilized for those patients with a marginal flow study would be that the decision was made by integrating the data from the intraoperative functional study and the preoperative anatomic findings.

In terms of accurate predictability, a clear decision point at a mean of ≈ 30 mm Hg is useful but not absolute. Carotti et al.⁸ showed that 7% (2/29) patients who had VSD closure based on these flow study parameters required subsequent fenestration of the VSD. We had successful late physiological outcomes (mid-term follow-up RVSP/SBP $<50\%$) at a flow study mean PA pressure of 30 mm Hg. We also had early failure with a mean PA flow study pressure of 25 mm Hg. The data available suggest that there is a marginal zone of physiological tolerance at a flow study pressure of ≈ 30 mm Hg. In fact, there was 1 patient who had the VSD left open and subsequently had too much pulmonary blood flow, requiring early VSD closure.

Relevance of the Data

By necessity, 2-dimensional anatomic data have been the gold standard by which preoperative surgical decision-making has been made. Implicit in that concept is that imaging findings correlate with postoperative functional outcomes. Anatomic variability within the disease of PA/VSD/MAPCA's lies on a continuum within which binary decisions regarding VSD closures must be made. Data on anatomic correlation to postoperative functional findings are marginal at best. In this study, the predictive value of conventional anatomic findings for VSD closure was 33% to 42%. Because of the limitations of imputing functional outcomes from 2-dimensional data, functional studies have become increasingly important in decision-making for those patients at the margin of sensitivity for anatomic studies. For example, we recently showed that functional MRI-based flow studies allowed a biventricular repair in up to one-third of neonates with marginal-size left ventricles who might have otherwise undergone a single ventricle repair.¹³ Although not perfect, functional data increase the accuracy and compress the confines of the marginal zone for binary decisions. The error rate for acute tolerances of VSD closure in this study was 5.2% (1/19), and the statistical predictive value of the flow study was 83%.

Implications of the Data

The data from this series lend further credence to the concept that all infants having the diagnosis of PA/VSD/MAPCA's should undergo an attempt at complete unifocalization. Reddy et al⁹ demonstrated that all 8 (32%) of their patients who required VSD fenestration had TNPAI <200 mm²/m², although a number of patients with TNPAI less than that were able to have their VSD closed. Carotti et al⁷ demonstrated that 75% of their patients having a TNPAI <150 mm²/m² and a staged repair eventually underwent VSD closure. In our experience, 9 out of 10 patients with TNPAI <150 mm²/m² underwent complete unifocalization and VSD closure. Said another way, once unifocalization had occurred, the functional study demonstrated potential for successful repair in 90% who would have been determined to be poor candidates for VSD closure based on anatomic findings.

MRI and Angiography-Based Functional Assessment

The intrinsic challenge of this method is that the flow study can be achieved only after major operative intervention. The study also occurs under nonpulsatile blood flow provided by CPB. Noninvasive preoperative assessment such as MRI may be very useful. In particular, MRI can provide both morphological and functional flow data. Flow and size of the MAPCA's and native PA's can thus be identified.¹⁴ A pulmonary-to-systemic blood flow ratio can be measured. One can also determine total indexed lung blood flow by measuring pulmonary vein flow, even in a typical-size neonate. Preoperative cardiac catheterization assessment with the mean pressure measurement in the MAPCA's combined with an MRI flow analysis could provide useful information regarding total pulmonary vascular resistance. This may become a means by which a preoperative functional predictor could replace an intraoperative assessment. The utility of preoperative functional assessment is currently being explored.

Conclusions

The present study demonstrates that the functional intraoperative pulmonary blood flow study is highly correlated with the postoperative RVSP and RVSP/SBP ratio and more accurately predicts successful VSD closure than anatomic indicators. Decisions regarding probability for successful complete repair should not be made on anatomic data alone. The implications of the findings are that all newborns having PA/VSD/MAPCA's should undergo a treatment algorithm that allows a functional test of their reconstructed pulmonary vasculature.

Acknowledgment

The authors thank Nobuko Yamamoto for her illustration.

Disclosures

K.D.T. was supported by a summer internship scholarship from the American Association for Thoracic Surgery.

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Circulation. 2009;120:S46-S52

doi: 10.1161/CIRCULATIONAHA.108.844084

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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