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Early Complete Repair of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collaterals

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Background. Pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) is a complex and diverse form of congenital heart defect. Although most patients with PA/VSD/MAPCAs can wait until they are 3 to 6 months of age to undergo surgical reconstruction, there are three specific criteria that merit an earlier repair. These 3 criteria are (1) unremitting heart failure; (2) a ductus to one lung and MAPCAs to the other; and (3) hemitruncus to one lung and MAPCAs to the other. The purpose of this study was to evaluate our surgical experience with early complete repair of PA/VSD/MAPCAs.

Methods. This was a retrospective review of patients undergoing complete repair of PA/VSD/MAPCAs within the first 60 days of life. Twenty-seven patients were identified in our database (2002 to 2013) who met these criteria. Fifteen had congestive heart failure, 9 had a ductus plus MAPCAs, and 3 had hemitruncus plus MAPCAs. The median age at surgery was 5 weeks.

Results. There was no operative mortality in this cohort of 27 patients. Hemodynamics at the conclusion of

the complete repair demonstrated an average right ventricular peak systolic pressure of 32 ± 5 mm Hg and an average right ventricle to aortic pressure ratio of 0.36 ± 0.06 . The median length of hospital stay was 26 days. There have been 2 subsequent mortalities (7%), with a median follow-up duration of 4 years. Eight of the 27 patients have subsequently undergone conduit replacements at our institution. The hemodynamics at the conclusion of the conduit change were statistically unchanged compared with the hemodynamics after complete repair.

Conclusions. The data demonstrate that early complete repair of PA/VSD/MAPCAs can be accomplished with low mortality and excellent postoperative hemodynamics. These early hemodynamic results are maintained at medium-term follow-up. We conclude that early complete repair is an appropriate choice for this highly select subgroup of patients.

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Pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) is a complex form of congenital heart disease. Advances in the treatment of PA/VSD/MAPCAs over the past 2 decades have resulted in a significant improvement in the prognosis for these patients [1, 2]. This experience has also highlighted the anatomic and physiologic diversity of this diagnosis and has revealed the need for a sophisticated management algorithm [3]. A midline unifocalization approach is applicable to the majority of patients with PA/VSD/MAPCAs, and thus provides the

mainstay of treatment [4–8]. However, there are also subsets of patients for whom alternative strategies must be employed [9–12]. There is growing evidence to suggest that some anatomic subsets will have a more or less favorable prognosis [13].

We routinely perform neonatal cardiac catheterization in patients identified with having PA/VSD/MAPCAs to define the anatomy and sources of pulmonary blood flow. The majority of patients can then safely wait until infancy (3 to 6 months) for their complete repair. Patients with the combination of cyanosis and dual-supply MAPCAs may be candidates for creation of an aortopulmonary window [14]. Conversely, there are three specific situations that merit an earlier repair to remedy excessive flow and pressure to the lungs; these are (1) unremitting heart failure; (2) a ductus supplying one lung and MAPCAs to the contralateral lung; and (3) hemitruncus to one lung and MAPCAs to the other. These criteria (one clinical

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and two identified by cardiac catheterization) define a subset of patients with PA/VSD/MAPCAs for whom we have recommended surgical repair within the first 60 days of life.

Although we believe that early complete repair is indicated for these three specific criteria, we have not previously studied this part of our management algorithm. The purpose of the present study was to summarize our experience with early complete repair for PA/VSD/MAPCAs.

Material and Methods

This study was approved by the Institutional Review Board at Stanford University. Patients were identified through the cardiac database, and the medical records were subsequently reviewed. The current study summarizes our experience with 27 patients, 16 male and 11 female, who were born with PA/VSD/MAPCAs and who met the criteria for early complete repair between the years 2002 and 2013. During this same period, there have been a total of 550 patients who received surgical care for the diagnosis of PA/VSD/MAPCAs at our center. Thus, the number of patients who underwent early complete repair represents approximately 5% of the entire cohort. Fifteen patients met our criteria of unremitting heart failure despite aggressive anticongestive medical therapy. These patients invariably demonstrated the signs and symptoms of pulmonary overcirculation, including tachypnea, poor feeding, and poor weight gain. These clinical symptoms were frequently associated with the need for hospitalization on a recurrent basis. The average ratio of pulmonary to systemic blood flow (as determined at cardiac catheterization) was 3.1 ± 0.7 .

Nine patients had the anatomic finding of a ductus to one lung (all 9 to the left lung, 7 with left aortic arches and 2 with right aortic arches) and MAPCAs to the contralateral lung (Fig 1). Many of these patients demonstrated clinical signs and symptoms of congestive heart failure, and could have met the criteria for early repair on the basis of unremitting heart failure had they not met the anatomic criteria identified by cardiac catheterization. The final subgroup of 3 patients had hemitruncus to one lung (2 the left lung, 1 to the right) and MAPCAs to the other (Fig 2). This small subgroup of patients with hemitruncus and MAPCAs was similar to the 9 patients with a ductus and MAPCAs from a clinical and physiologic basis. The average age at surgery was 34 ± 7 days (range, 7 to 60), and the average weight at surgery was 4.4 ± 0.6 kg (range, 2.1 to 5.6 kg). Twelve of the 27 patients were hospitalized before surgery owing to their tenuous clinical situation.

Figure 3 demonstrates an illustration of the surgical technique utilized for unifocalization of a ductus or hemitruncus to one lung with MAPCAs to the contralateral side. We do not perform an intraoperative flow study as these patients have very well developed pulmonary vascular beds with low pulmonary vascular resistance. Figure 4 shows an illustration of the completed repair using an aortic homograft from the

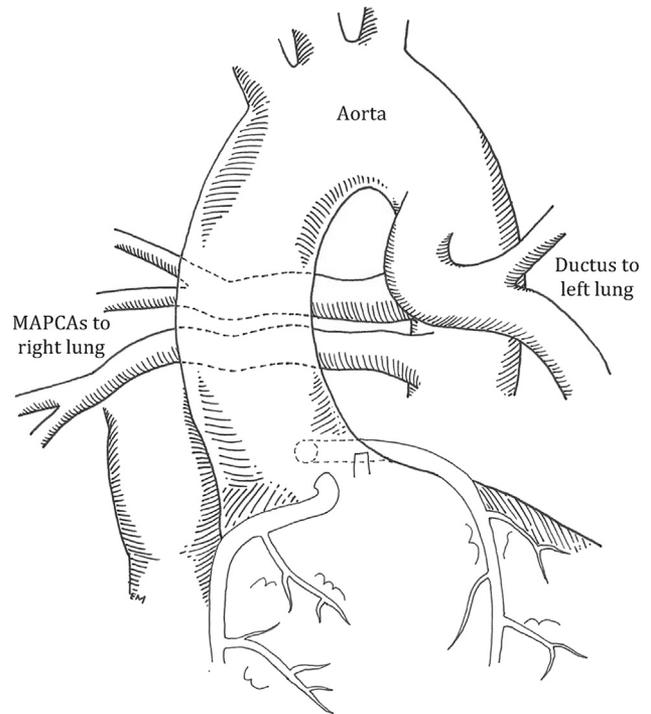


Fig 1. The left pulmonary artery is shown originating from a left-sided ductus and supplying the entire left lung. The right lung is supplied by two major aortopulmonary collaterals. (MAPCAs = major aortopulmonary collaterals.)

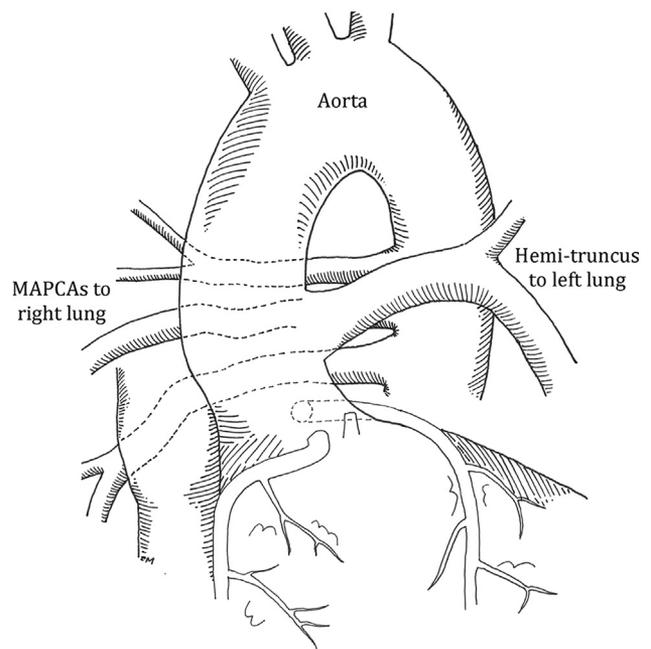


Fig 2. The left pulmonary artery is shown originating from the ascending aorta (hemitruncus). There are three major aortopulmonary collaterals that supply the right lung. (MAPCAs = major aortopulmonary collaterals.)

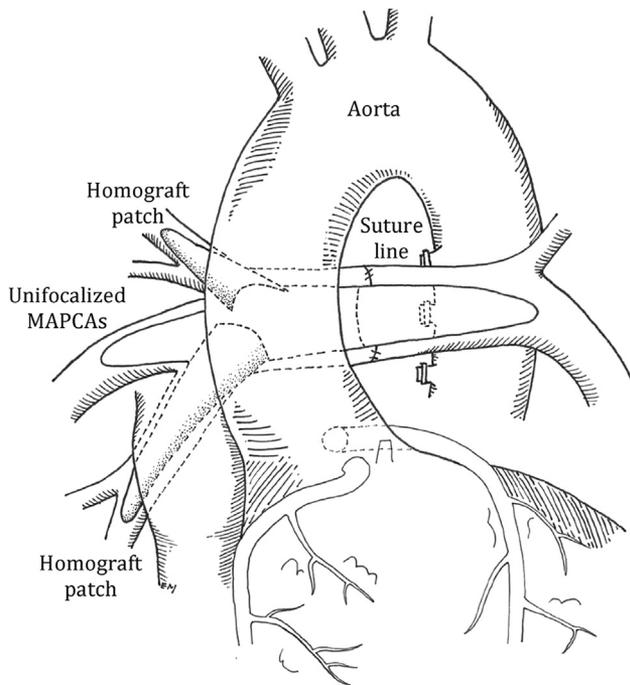


Fig 3. The technique utilized for unifocalization of a ductus or hemitruncus to one lung and major aortopulmonary collaterals (MAPCAs) to the contralateral lung is illustrated. The ductus or hemitruncus is harvested from the aorta, and the site of origin is oversewn. The MAPCAs are harvested from the descending aorta and are brought together to form a confluence. Each of the three MAPCAs is augmented with pulmonary artery homograft. The posterior wall of the ductus/hemitruncus is sutured to the posterior wall of the MAPCA confluence, providing a living tissue to living tissue connection.

right ventricle to pulmonary artery. We have utilized aortic homografts for these reconstructions because the unifocalized MAPCAs result in pulmonary artery pressures that are modestly elevated compared with a normal pulmonary vascular bed. It has been our belief that aortic homografts provide a more durable conduit, particularly with respect to limiting the degree of conduit insufficiency. A summary of the homograft conduit sizes utilized to restore continuity from the right ventricle to reconstructed pulmonary arteries is presented in [Table 1](#). These homograft sizes were selected based on a combination of patient size, coronary anatomy, and homograft availability. At the conclusion of the complete repair, left atrial and right ventricular pressures were monitored by placement of two transthoracic lines. The right ventricle peak systolic pressure and aortic peak systolic pressure were recorded after separation from cardiopulmonary bypass. These pressure measurements were recorded in the operative notes as well as the calculation of right ventricle to aortic pressure ratio. Eight of the 27 patients have undergone conduit replacements at our institution. The average interval between complete repair and conduit change was 2.5 ± 1.0 years.

During the time frame of this study, there were 3 patients who met the anatomic criteria for early complete

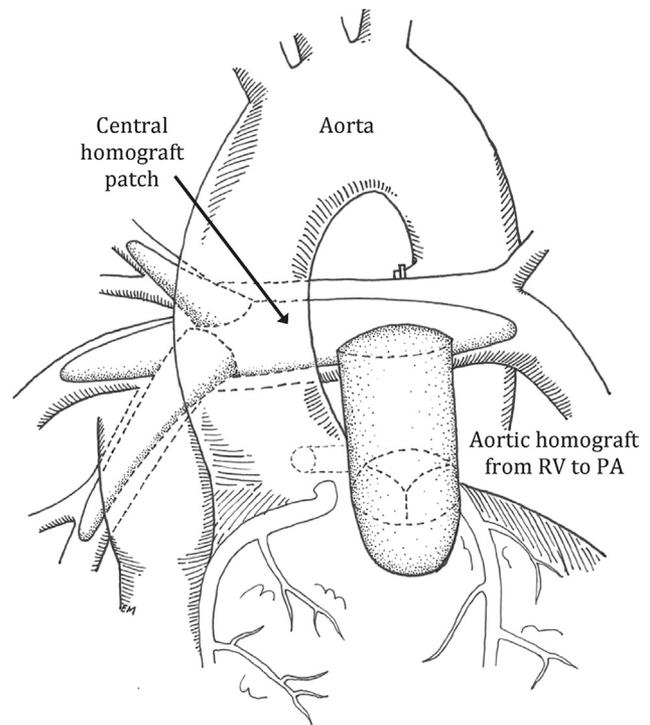


Fig 4. A complete repair of a ductus or hemitruncus to the left lung and major aortopulmonary collaterals to the right lung is illustrated. A homograft patch has been used to augment the central “pulmonary arteries.” An aortic homograft is placed to establish continuity from the right ventricle (RV) to the pulmonary arteries (PA).

repair but were not considered to be suitable candidates for a single-stage approach. All 3 of these patients had a ductus to the left lung with small MAPCAs to the right lung and a relatively underdeveloped right pulmonary vascular bed. These 3 patients underwent a staged surgical approach, and ultimately, all were able to achieve complete repair. Statistical results are reported as the mean \pm SD. Comparison of different temporal points was performed with a paired *t* test analysis.

Results

There was no operative mortality in this cohort of 27 patients undergoing early repair of PA/VSD/MAPCAs. All 27 were able to have a complete single-stage procedure. At the conclusion of this procedure, the average right ventricular peak systolic pressure was 32 ± 6 mm Hg with an average aortic peak systolic pressure of 90 ± 18 mm Hg. That would result in an average right ventricle to aortic peak systolic pressure ratio of 0.36 ± 0.07 . Calculation of the pulmonary vascular resistance revealed an average of 2.0 ± 0.30 Woods units. The average duration of hospital stay from the date of operation to date of discharge was 31 ± 8 days (range, of 9 to 106; median 26). There were three postoperative complications that contributed to an increase in hospital stay, including wound infection in 1 patient, pneumonia in 1, and chylothorax in 1. The most common reason for

Table 1. Conduit Sizes in Patients Undergoing Early Complete Repair of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collaterals

Conduit Size	Patients
8 mm	n = 4
9 mm	n = 5
10 mm	n = 5
11 mm	n = 6
12 mm	n = 5
13 mm	n = 1
14 mm	n = 1

prolonged stay was feeding issues and establishing adequate weight gain.

All 27 patients were eventually discharged from the hospital after their surgical repair. The patients have been followed up for an average of 4 ± 1 years. There have been 2 deaths (7%) during the follow-up period, and these occurred 3 and 4 months after repair. The first patient had pneumonia and sepsis. The second patient underwent complete repair of a ductus to the left lung and relatively small MAPCAs to the right lung. The post-operative right ventricular pressures were half systemic, but a subsequent lung perfusion study demonstrated that more than 90% of pulmonary blood flow was to the left lung. This patient underwent a series of attempted palliations, but eventually died of right heart failure.

Eight of the 27 patients have undergone conduit replacement at our institution. The indication for conduit replacement was severe stenosis in all 8 patients. After conduit change, the average right ventricular peak systolic pressure was 32 ± 5 mm Hg and the simultaneous aortic peak systolic pressure was 96 ± 8 mm Hg. Therefore, the calculated right ventricle to aortic pressure ratio after conduit replacement was 0.34 ± 0.05 (Fig 5). The pressure ratios after complete repair and after conduit replacement were not statistically different.

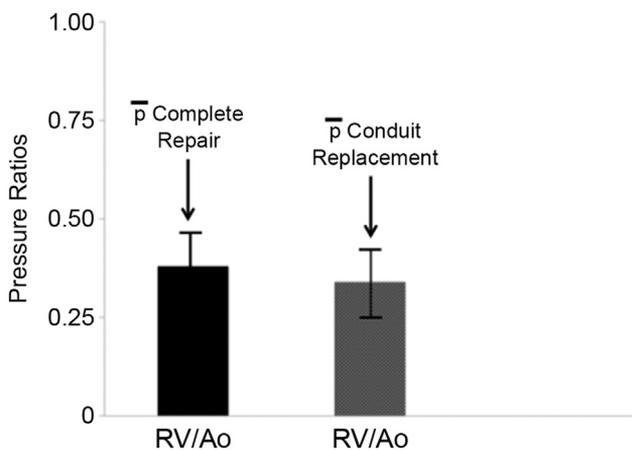


Fig 5. A bar graph demonstrates the peak systolic right ventricle (RV) to aortic (Ao) pressure ratios at the conclusion of the complete repair and after conduit replacement.

Comment

Herein, we have summarized our experience in 27 patients who underwent early complete repair of PA/VSD/MAPCAs. The criteria for recommending early complete repair (less than 60 days of age) included unremitting heart failure, a ductus to one lung and MAPCAs to the contralateral lung, or hemitruncus to one lung and MAPCAs to the other lung. The results of this study demonstrated no operative mortality and a relatively low interim mortality rate. The average right ventricle to aortic peak systolic pressure ratio was 0.36 ± 0.07 after early complete repair. These findings suggest that the strategy of early complete repair is appropriate for this select subset of patients with PA/VSD/MAPCAs.

Unremitting heart failure in neonates with PA/VSD/MAPCAs is invariably associated with large, unrestrictive MAPCAs that result in excessive pulmonary flow. These patients also have a degree of cyanosis, and the combination would seem to be more problematic than pulmonary overcirculation alone [15]. From a clinical standpoint, these patients are similar to neonates with other complex forms of congenital heart disease. The closest example of this is in patients with truncus arteriosus, who have this same combination of pulmonary overcirculation and cyanosis. Efforts at medical management are frequently not productive in achieving the goals of symptomatic improvement, better feeding, and weight gain. In addition, medical management may be fraught with hazards, as these fragile neonates are at risk for respiratory and infectious complications, and this strategy has been associated with a high attrition rate. As a consequence, most patients with complex congenital heart disease and unremitting heart failure are best served through early anatomic repair [15, 16].

These historical lessons are equally applicable to PA/VSD/MAPCAs, a lesion for which there is no viable option for intermediate palliation. It is acknowledged that we do not know specifically how this cohort of patients would have done with ongoing medical management, and there is no randomized, prospective study comparing medical versus early surgical treatment in this setting. We would suggest that such a study is not indicated, as it would recapitulate the lessons learned several decades ago that led to the adoption of a more aggressive surgical time schedule. The two anatomic diagnoses that we have included as criteria for early repair are both identified at the time of neonatal cardiac catheterization. Although it is evident that the anatomic sites of origin for a ductus and hemitruncus are not identical, from a physiologic standpoint, these two anatomic variants are similar in that they result in systemic level pressure to the supplied lung. The combination of systemic level pressure and excessive pulmonary blood flow can lead to the early development of pulmonary vascular obstructive disease [17]. Our early experience with these two anatomic entities indicated the importance of identifying and repairing them before the onset of irreversible changes.

The single-stage complete repair rate for this cohort of 27 patients was 100%, which compares quite favorably to

our overall experience with PA/VSD/MAPCAs [1–3]. We have previously published a single-stage complete repair rate of 55% for all nonselected patients presenting to our institution with this diagnosis. This disparity in complete repair rate may at first appear surprising, given the clinical acuity and relatively nonelective nature of the early repair candidates. However, patients with PA/VSD/MAPCAs who manifest heart failure have an extremely well developed pulmonary vascular bed with low pulmonary vascular resistance. A low pulmonary resistance is the physiologic reason why heart failure develops in these patients, but for the same reason is also a reliable marker for success of a single-stage operation. In the hierarchy of suitability for single stage repair, patients with a balanced circulation (Qp:Qs close to 1) tend to have moderate development of their pulmonary vascular bed and are consequently the next most suitable candidates. Finally, patients who present with cyanosis tend to have a poorly developed pulmonary vascular bed, have a low rate of single-stage complete repair, and probably have a worse short-term and long-term prognosis [13].

The majority of patients with PA/VSD/MAPCAs who we manage at our center undergo surgical intervention between 3 and 6 months of age. We have found this timeline usually permits some modest visceral growth and maturation of the lung parenchyma. It is noteworthy that the MAPCAs will frequently undergo changes between the neonatal cardiac catheterization and the one performed several months later. The most commonly observed changes observed are proximal stenosis close to the origin of the MAPCAs from the aorta and stenosis developing at the bifurcation (or trifurcation) of the lobar or segmental branches. These stenoses may progress to complete occlusions if surgical intervention is delayed too long. When occlusions do occur, it typically results in the loss of those segments of lung supplied by that vessel. Therefore, the time constraints for performing surgical repair are dictated by the natural course of history for MAPCAs.

Although we have outlined the rationale for advancing the timeline of intervention for patients with PA/VSD/MAPCAs and unremitting heart failure or a ductus/hemitruncus to one lung and MAPCAs to the other, there are several disadvantages to early complete repair. The most notable is the fact that these neonates or young infants, many of whom are in profound heart failure, typically require prolonged hospital stays for their convalescence. In this series, the average length of stay after surgery was 31 days, which is nearly twice as long as the lengths of stay for the “standard” infant undergoing complete repair [3]. Much of this additional convalescence was dedicated to the management of feeding issues and achieving the goal of weight gain. This clinical observation is not surprising given the difficulty of achieving these goals in neonates and young infants with profound heart failure. The other drawback to early repair is that the sizes of the right ventricle to pulmonary artery conduits are obligatorily small, and thus are outgrown at a relatively early age. The median conduit size in this series was 11 mm, and it is evident that the longevity of small conduit size is quite limited.

The 27 patients have been followed for an average of 4 years. During this follow-up period, 8 patients have undergone conduit replacement at our institution. In these 8 patients, the right ventricle to aortic pressure ratio was slightly lower at the end of the conduit replacement (0.34) than compared with the end of the complete repair (0.36), recognizing that there was no statistical difference given the small number of patients. These hemodynamic numbers are comparable to those previously reported for patients undergoing elective repair [18]. We are aware of 8 additional patients who have undergone replacement of their right ventricle to pulmonary conduits at outside institutions. We receive patient referrals from a large geographic area for the unifocalization procedure, but most institutions are comfortable performing their own conduit changes. As a consequence, we do not have comprehensive hemodynamic data for these patients and have limited access due to the consequences of the Health Insurance Portability and Accountability Act (HIPAA).

There were 3 patients who met the anatomic criteria but were not considered to be suitable for a single-stage complete repair. These patients had a ductus to the left lung with relatively small MAPCAs to the right lung. Our past experience showed us that unifocalization in the circumstance of markedly disparate blood flow results in nearly all of the pulmonary blood flow going to the well-developed lung. Our algorithm for evaluating cases of ductus or hemitruncus to one lung and MAPCAs to the other lung is based on an assessment of the relative pulmonary blood flow to each lung. We would proceed with complete repair as long as the disparity in relative blood flow does not exceed twofold. For patients who exceed this limit, we would recommend unifocalization of the MAPCAs to a central shunt while maintaining discontinuity with the left lung. In this circumstance, if the ductus shows signs of narrowing, we would proceed with placement of a ductal stent. It is likely that one of the deaths in this series was the result of proceeding with complete repair in a patient who had underdeveloped MAPCAs to the contralateral lung. The 3 patients who had small MAPCAs to the right lung and who were managed in a stepwise fashion have subsequently undergone reconstruction of the right and left pulmonary arteries with complete repair.

In summary, the results of this study demonstrate that patients with PA/VSD/MAPCAs who underwent early complete repair had excellent short-term outcomes. The postoperative right ventricle to aortic pressure ratios were comparable to previously published data for patients undergoing elective repair. The midterm hemodynamic data demonstrate that right ventricle to aortic pressure ratios are maintained over time, implying the unifocalized pulmonary vascular bed has the capability to keep pace with visceral growth. These results indicate that early complete repair is an appropriate strategy for this select subgroup of patients.

Figures 1 through 4 were illustrated by Erin Anne Mainwaring.

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DISCUSSION

DR JOHN H. CALHOON (San Antonio, TX): Dr Watanabe, what a beautifully presented paper and very clear slides. Thank you very much for sending me the manuscript in advance.

I think your paper and your premise points out to me something that's kind of always irrefutable, and that is that we have to pay attention to the symptoms and the anatomy, and what you have described out of some 500 patients was about 5% of them who presented with either unremitting heart failure or a chance to get pulmonary vascular obstructive disease. You selected those patients out and found that you could operate on them early and get great results, as you have shown in other of these patients.

You had 3 patients in whom you noted were the major aortopulmonary collaterals (MAPCAs) were "too small" on one side. I wonder if you could share with us what you learned in this experience and how you might decide if a MAPCA is too small.

You also in your illustration pointed out you used an aortic (Ao) homograft. Do you always use an aortic homograft, do you always, I'm assuming, use a valved homograft, and do you have a particular preference for the reconstruction of the right ventricular outflow tract? Congratulations once again on a beautifully presented and well written paper. Thank you for the chance to discuss it.

DR WATANABE: Thank you for the questions. To answer the first question, we evaluate whether a patient can have a primary versus

secondary repair based on the relative distribution of pulmonary blood flow on cardiac catheterization. If the angiogram demonstrates that one lung receives more than two times the other lung, we would not perform an early complete repair. Instead, we would unifocalize the MAPCAs to a shunt and maintain discontinuity to the contralateral side. We would wait 3 to 6 months and at that time bring the right and left sides together.

And for the second question, we use aortic homografts for the right ventricle (RV) to pulmonary artery (PA) conduit. One of my slides showed the RV/Ao pressure is between 30% and 40% systemic, which is higher than normal. We want to avoid conduit insufficiency and aneurysms of the conduit, and that is the reason why we choose aortic homografts.

DR JOHN E. MAYER (Boston, MA): Just two short questions. One is, could you tell us anything about the reintervention rate in terms of interventional catheterization in this subset of patients? And, number two, do you have any lung scan data in follow-up to give us an idea about the distribution of blood flow between your MAPCA lung and your ductal-dependent lung, if you will? Congratulations on a nice series.

DR WATANABE: Thank you very much. We usually do a lung perfusion scan before discharge, but I don't have the results of those data now. So I'm not sure of the number.

DR MAYER: I was just interested in the intervention or reintervention rate, presumably mostly by catheterization, for each of these patients.

DR WATANABE: We have performed eight conduit replacements after the neonatal complete repair. All of these patients undergo repeat cardiac catheterization to reassess the anatomy and physiology before conduit change. But this represents only one third of the patients.

DR RICHARD MAINWARING: John, let me add a note of clarification. Obviously one of the shortcomings of this approach is that the babies are young and end up with small conduits. When these patients come back for their conduit changes, we have an opportunity to evaluate the reconstructed pulmonary anatomy. If we see a stenosis in one of the pulmonary branches, we would prefer to repair this surgically at the time of conduit change. We rarely use interventional techniques for this purpose.

DR JORGE D. SALAZAR (Jackson, MS): For our higher risk patients with MAPCAs that are questionable in size, we have performed fenestrated ventricular septal defect (VSD) closures in concert with an RV to PA conduit. What is your experience with this approach?

DR WATANABE: We do not use fenestrations of the VSD patch. We perform an intraoperative flow study, and if this study shows good numbers, we close the VSD. If the flow study does not have acceptable numbers, then we do not close the VSD at that surgery.

DR SALAZAR: And if you don't close the VSD when you unifocalize, do you unifocalize to a shunt or do you unifocalize to an RV to PA Sano-like conduit?

DR WATANABE: We do a central shunt.