Late Outcomes in Patients Undergoing Aortopulmonary Window for Pulmonary Atresia/Stenosis and Major Aortopulmonary Collaterals

Richard D. Mainwaring, MD, V. Mohan Reddy, MD, Stanton B. Perry, MD, Lynn Peng, MD, and Frank L. Hanley, MD

Divisions of Pediatric Cardiac Surgery and Pediatric Cardiology, Lucile Packard Children's Hospital at Stanford University, Palo Alto, California

Background. Pulmonary atresia with ventricular septal defect (PA/VSD) and major aortopulmonary collateral arteries (MAPCAs) is a complex form of congenital heart defect. One identifiable subset has small (<2.5 mm) intrapericardial branch pulmonary arteries that are (1) confluent, (2) have normal arborization, and (3) have dual-supplied collateral vessels. When this anatomy is associated with limited pulmonary blood flow, the patients are candidates for creation of an aortopulmonary window to stimulate growth of the pulmonary arteries. The purpose of this study was to review our experience with creation of an aortopulmonary window as the initial palliative procedure.

Methods. This was a retrospective review of our surgical experience with 35 children undergoing aortopulmonary window creation from 2002 to 2011. Patients were identified by preoperative cardiac catheterization to define the cardiac and pulmonary artery anatomy.

Results. There was no mortality in 35 patients undergoing aortopulmonary window creation. These patients have subsequently undergone 78 cardiac procedures (with 2 operative mortalities). Eighteen of these patients have achieved complete repair, 4 patients in a second procedure, 6 patients in a third procedure, 5 patients in a fourth procedure, and 3 patients in a fifth procedure.

Conclusions. The data demonstrate that patients can undergo creation of an aortopulmonary window with excellent early results. Few patients were amenable to complete repair at the second operation, and most required multiple reoperations to recruit sufficient arborization. We interpret these counterintuitive results to suggest that hypoplastic central pulmonary arteries and diminished pulmonary blood flow are markers for a less well developed pulmonary vascular bed.

(Ann Thorac Surg 2012;94:842–9)
© 2012 by The Society of Thoracic Surgeons

Pulmonary atresia with ventricular septal defect (PA/VSD) and major aortopulmonary collateral arteries (MAPCAs) is a highly variable form of congenital heart disease [1]. The majority of patients with MAPCAs have abnormal arborization of the pulmonary arteries or a complete absence of intrapericardial pulmonary arteries [2]. These patients are managed surgically through the "unifocalization" of MAPCAs into a central confluence to serve as the predominant source of pulmonary blood flow [3]. The mid- and late-term outcomes for patients undergoing a midline unifocalization approach for PA/VSD MAPCAs have improved significantly over the past 15 years [4], and these favorable results have been confirmed by multiple groups [5–9]. There is now a majority consensus that this approach improves the prognosis for patients with PA/VSD MAPCAs compared with the untreated "natural history" [10, 11].

There exists a smaller subset of patients with MAPCAs who have diminutive intrapericardial pulmonary arteries that are confluent, have normal arborization, and provide "dual blood supply" with pulmonary segments that have blood flow from MAPCAs. In the clinical setting of cyanosis, this group of patients is amenable to the creation of an aortopulmonary window. This initial palliative approach frequently leads to growth of the entire native pulmonary artery system and may obviate the need for unifocalization of the MAPCAs. Although our group has championed the midline unifocalization strategy for the majority of patients with MAPCAs, we also recognize that there is a small subset of patients amenable to creation of an aortopulmonary window. We have previously outlined our guidelines and results for this specific group [12].

There is currently a paucity of data regarding the influence of specific anatomic features on outcomes. Several recent publications have found conflicting results with regard to the importance of pulmonary artery anatomy [6–8]. In addition, these studies did not evaluate the subgroup of patients with confluent pulmonary arteries and normal arborization. The purpose of the present study was to review our mid- to late-term outcomes with

Ann Thorac Surg
2012;94:842–9

MAINWARING ET AL
AORTOPULMONARY WINDOW

843

PEDIATRIC CARDIAC

Fig 1. (A) Angiogram demonstrating diminutive branch pulmonary arteries that are confluent, have normal arborization, and share dual blood supply with the aortopulmonary collateral arteries. (B) Postoperative angiogram of same patient shown in A, demonstrating growth of the native pulmonary arterial system.

aortopulmonary window creation as the initial palliative procedure.

Material and Methods

This study was approved by the Institutional Review Board at Stanford University. Patients undergoing creation of an aortopulmonary window were identified through the cardiac database. A questionnaire and Health Insurance Portability and Accountability Act authorization form were sent to the families. A returned questionnaire signified consent by the parents for review of the medical records.

The current study summarizes our surgical experience with 35 patients undergoing creation of an aortopulmonary window from April 2002 through December 2011. There were 19 boys and 16 girls included in this study. The median age at the time of operation was 5 weeks (range, 4 days–4 weeks). The median weight at operation was 4.4 kg (range, 2.1–5.6 kg).

A prerequisite for this procedure was cyanosis in conjunction with the anatomy of small (<2.5 mm) intrapericardial branch pulmonary arteries that were documented by cardiac catheterization to (1) be confluent, (2) have normal arborization, and (3) have dual-supplied multiple collateral arteries. This anatomy is demonstrated in the angiogram shown in Fig 1.

Detailed angiography was performed preoperatively in all patients to delineate the anatomy of the central branch pulmonary arteries and major aortopulmonary collaterals. Sixteen of the 35 patients had pulmonary atresia with dual supply to all lung segments. Nine patients had some antegrade flow through the pulmonary valve, and thus would be categorized anatomically as pulmonary stenosis. However, the amount of pulmonary blood flow was so minimal that these patients still fulfilled the criteria that we utilize for aortopulmonary window (ie, cyanosis, branch pulmonary arteries <2.5 mm, dual supply collaterals), and thus physiologically had "near-atresia" of the pulmonary valve. There were six patients who had pulmonary atresia with dual supply to most (between 13–17) but not all segments, and thus by definition had some

segments with single supply MAPCAs. Finally, there were four patients who had PA/VSD/MAPCAs in conjunction with other complex anatomy such as corrected transposition.

The surgical technique for creation of an aortopulmonary window is through a median sternotomy with cardiopulmonary bypass on standby. The intraoperative anatomy is inspected to corroborate the data obtained from the preoperative cardiac catheterization (Fig 2). The
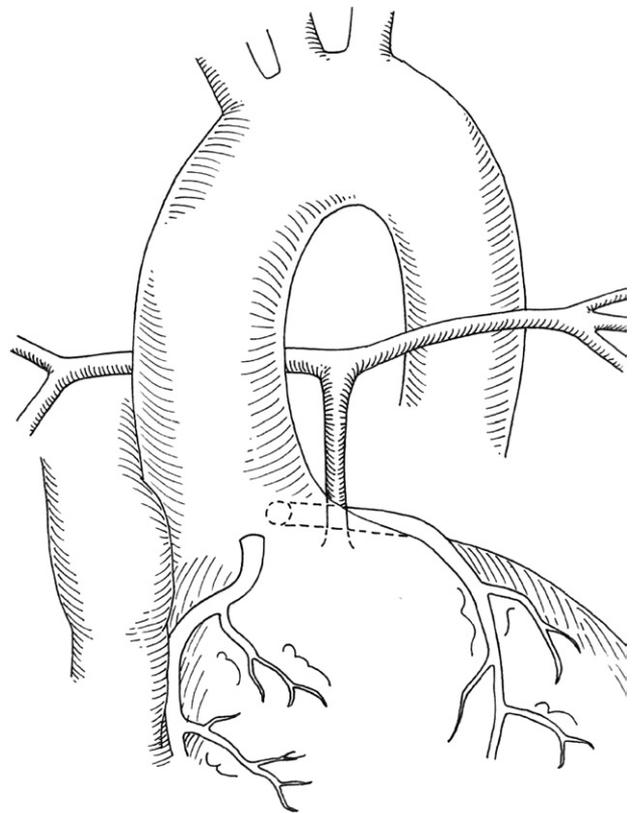

Fig 2. Anatomy in pulmonary atresia with confluent but hypoplastic pulmonary arteries. The branch pulmonary arteries measure 1 to 2 mm in diameter in this subgroup of patients.

Fig 3. Illustration of surgical technique used in creating an aortopulmonary window. The right and left branch pulmonary arteries have been fully mobilized and the atretic main pulmonary artery divided in preparation for the anastomosis. The site for reimplantation of the main pulmonary artery is denoted by the dashed oval along the leftward and posterior aspect of the aorta. The inset demonstrates the correct positioning of this site, which is at the 5 o'clock position to avoid kinking of the branch pulmonary arteries. (LMCA = left main coronary artery; RCA = right coronary artery.)
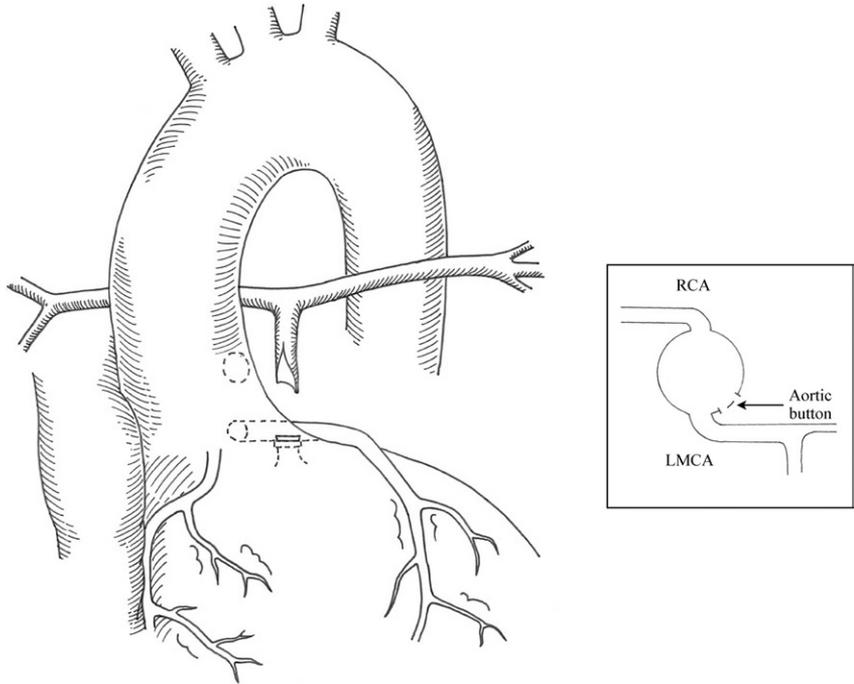
right and left branch pulmonary arteries are mobilized and occluded, and the atretic main pulmonary artery is divided. The location for the anastomosis on the aorta is quite posterior and is marked in advance with a suture (as illustrated in Fig 3). This posterior location is important to avoid potential kinking or stretching of the right branch pulmonary artery. A side-biting clamp is then placed on the left posterolateral aspect of the ascending aorta, and a small button of aorta is removed with a No. 11 blade (Fig 4). The main pulmonary artery segment is incised for a distance of 3 mm, and the anastomosis is performed with 7-0 or 8-0 polypropylene suture. It is important to maintain proper alignment of the diminutive main pulmonary artery, as shown in Fig 5. After completion of the aortopulmonary anastomosis, the side-biting clamp is removed. The observation of significant distention of the central branch pulmonary arteries, along with an increase in oxygen saturation, provides confirmation of a well-functioning aortopulmonary window. Direct stick pressures can be performed as further confirmation of a satisfactory anastomosis.

The advantages of the aortopulmonary window technique compared with a standard systemic-to-pulmonary artery shunt are numerous. First, the tissue-to-tissue anastomosis greatly reduces the risk of thrombosis. Second, the aortopulmonary window is less likely to result in distortion of the branch pulmonary arteries, which by definition are markedly diminutive at the time of this operation. Finally, the central location of the aortopulmonary window usually results in balanced blood flow distribution to the right and left branch pulmonary arteries. However there are rare patients who have confluent central branch pulmonary arteries exceeding our size limit of 2.5 mm, and under these circumstances we

would advocate using a central polytetrafluoroethylene shunt. This prosthetic shunt will regulate flow and pressure and lessen the possibility of excessive pulmonary blood flow or a reperfusion injury to the lung, or both. We do not have any experience with right ventricular outflow tract reconstruction in this setting as advocated by some [8] and thus cannot comment on the relative
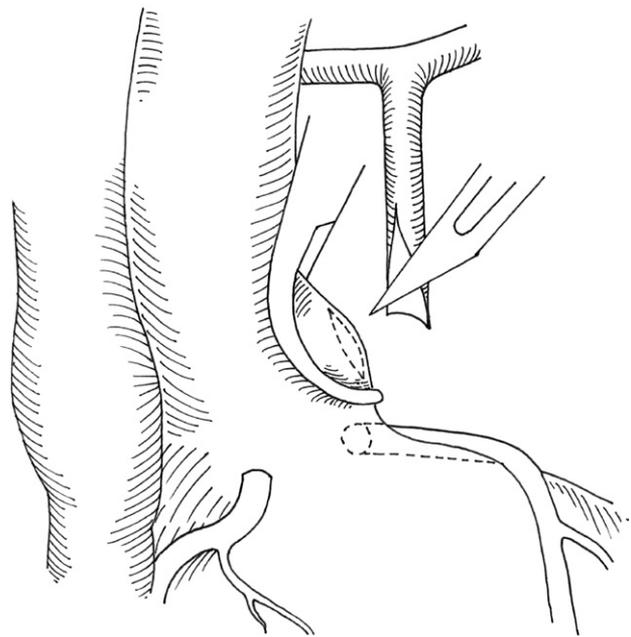

Fig 4. Preparations for the aortopulmonary window. A side-biting clamp has been placed on the posterolateral aspect of aorta, and a small button of the aortic wall (toward posterior aspect) is removed.
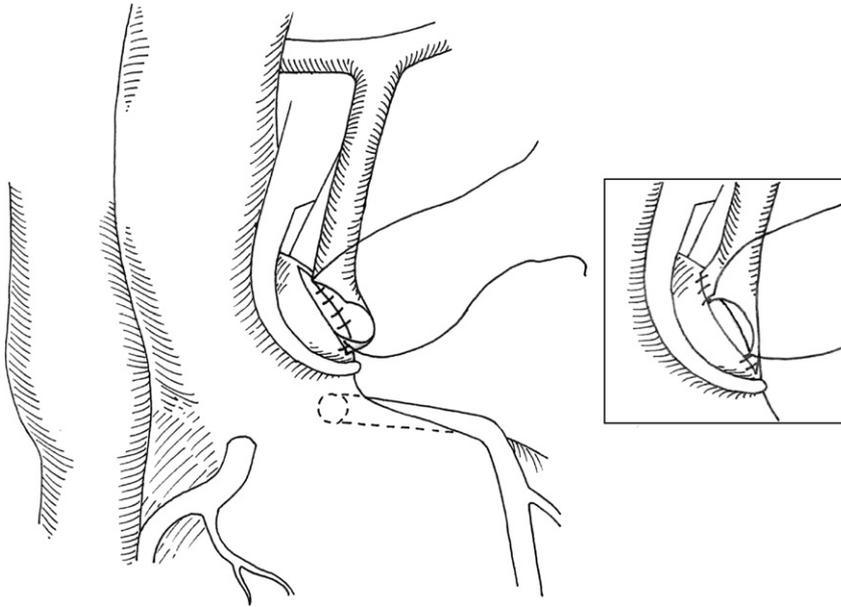
Fig 5. Anastomosis between diminutive main pulmonary artery and ascending aorta. It should be noted that correct positioning of "heel" of main pulmonary artery is at the 2 o'clock position on the aortotomy (as shown in the figure) and not top of the aortotomy. Anastomosis is performed with 7-0 or 8-0 monofilament absorbable suture.

merits of this approach. However our preference is to avoid conduits or outflow patches when they will be exposed to systemic pressure because of a significant incidence of false aneurysms in this circumstance.

Statistical analysis was performed using Fisher's exact test. The results of this study were compared with previously published data for patients undergoing a standard unifocalization approach. A p value less than 0.05 was considered statistically significant.

Results

There was no surgical mortality in this cohort of 35 patients undergoing creation of an aortopulmonary window. There were 3 major complications, including sepsis in 2 patients and a cardiac arrest requiring cardiopulmonary resuscitation in 1 patient. There were 8 minor complications, including wound infection in 3 patients, respiratory complications requiring reintubation in 2 patients, prolonged pleural effusions in 2 patients, and transient renal failure in 1 patient. The median duration of hospital stay after operation was 16 days (range, 7–64 days).

The patients have been followed for a median of 49 months (range 1–114 months). The 2 most recent patients

in this series did not have any further treatment after creation of the aortopulmonary window. The remaining 33 patients underwent cardiac catheterization at 4 months after the initial operation, demonstrating growth of the branch pulmonary arteries in all patients, with a mean pulmonary artery pressure of 53 mm Hg (median range, 38–72 mm Hg). These 33 patients underwent the second cardiac procedure at an average of 5.5 ± 1.0 months after creation of the aortopulmonary window, and these procedures are summarized in Table 1. There was 1 operative mortality among the 33 patients undergoing the second cardiac procedure. This occurred in an infant who had concomitant repair of both total anomalous pulmonary venous connection and moderate to severe atrioventricular valve regurgitation.

A summary of the subsequent 45 cardiac procedures in the 32 surviving patients is shown in Table 2. There was 1 operative mortality in a patient undergoing the third cardiac procedure, which was a revision of the previous unifocalization procedure plus a central shunt. There have been 3 late deaths unrelated to surgical procedures. The actuarial survival curve for the 35 patients undergoing creation of an aortopulmonary window is shown in Fig 6. The survival was 87% at the 4-year mark after creation of the aortopulmonary window.

A total of 18 of the 33 eligible patients (55%) have achieved complete repair, including closure of all intra-

Table 1. Summary of the 33 Procedures Performed as the Second Operation After Initial Aortopulmonary Window Creation

Complete single-stage repair	n = 4
Complete unifocalization + shunt	n = 9
Unilateral unifocalization	n = 16
Augmentation of central confluence	n = 3
Repair of TAPVC/repair of AV valve	n = 1

AV = atrioventricular;   TAPVC = total anomalous pulmonary venous connection.

Table 2. Summary of the 45 Cardiac Procedures Performed as the Third (or Greater) Operation Performed in the 32 Surviving Patients

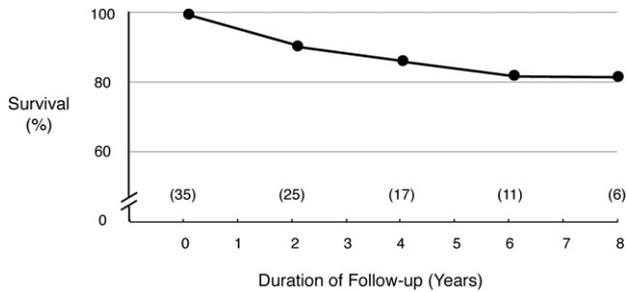|	
Complete repair	n = 14
Bilateral unifocalization revision	n = 13
Unilateral unifocalization revision	n = 11
Conduit change	n = 7

Fig 6. Actuarial analysis of 35 patients undergoing aortopulmonary window.

cardiac shunts and placement of a conduit from the right ventricle to the pulmonary artery. Four of the 33 patients were able to achieve complete repair at the second procedure. An additional 6 patients had complete repair at the third procedure, 5 patients at the fourth procedure, and 3 patients at the fifth procedure. There are 2 patients who have not undergone their second surgical procedure yet, and 7 additional patients who have not undergone repair but may be eligible for repair sometime in the future.

There were 5 patients (14%) who underwent ligation of MAPCAs at the time of creation of the aortopulmonary window. Ligation was performed on the basis of a large MAPCA identified by catheterization that potentially would provide excessive competitive pulmonary blood flow. There were also 16 patients (49%) who subsequently had incorporation of MAPCAs through standard unifocalization techniques. This was performed in the presence of important peripheral stenoses in the native pulmonary arterial system. By definition, these segments derived dual blood supply through MAPCAs, and under these circumstances it was technically easier to perform

unifocalization on the MAPCA than to repair the peripheral pulmonary artery stenoses. Twelve patients had ligation of MAPCAs that provided competitive flow to segments of the lung that were already supplied by unobstructed branches of the native pulmonary arterial system.

Seven patients have subsequently undergone conduit replacement after complete repair. All 7 of these patients had cardiac catheterization performed before conduit change. This cardiac catheterization provided an opportunity to assess complete hemodynamics in this limited subset of patients. The median pulmonary artery–to-aorta systolic pressure ratio was 0.39, with a range of 0.30 to 0.54.

Comment

This article summarizes our surgical experience with 35 patients who underwent creation of an aortopulmonary window as their initial palliation for PA/VSD and MAPCAs. There was no surgical mortality at the time of aortopulmonary window creation. These patients have subsequently undergone 78 additional cardiac procedures (for a total of 113 procedures), with 2 operative and 3 late nonoperative mortalities. Twelve percent of the patients were amenable for complete repair at the second surgery, and 55% of the patients have ultimately achieved complete repair. These results indicate that creation of an aortopulmonary window is a very safe early approach but typically requires multiple operations to recruit sufficient pulmonary arborization to allow septation of the heart.

We have previously published our institutional experience demonstrating that 88% of patients with PA/VSD MAPCAs derive the majority of their pulmonary blood flood from MAPCAs (Fig 7). These patients have either

Fig 7. Algorithm for surgical management of patients with pulmonary atresia (PA), ventricular septal defect (VSD), and major aortopulmonary collateral arteries (MAPCAs). Percentages shown are the proportion of total patients in the authors' experience. (AP = aortopulmonary.) (Reprinted from Malhotra SP and Hanley FL, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009;12:145-51 [3], with permission from Elsevier.)
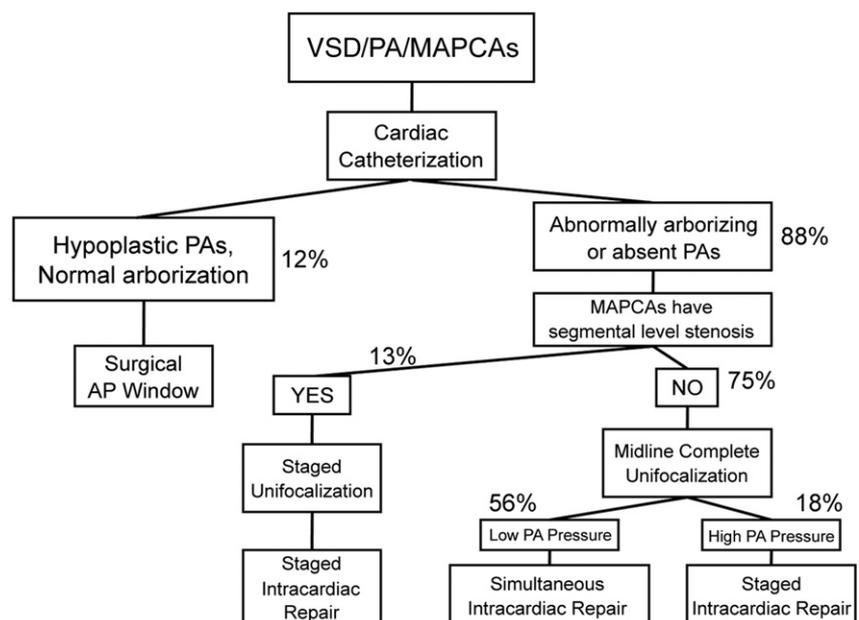
Table 3. Comparison of Group Undergoing Aortopulmonary Window Creation to Previously Published Series of Patients Undergoing Unifocalization of MAPCAs

Study Parameter	AP Window (n = 33)	Published Series (n = 462)
Single-stage complete repair	12%[a]	64%
Ultimate complete repair	55%[a]	90%
Alive at 4 years' follow-up	87%	92%

[a] $p < 0.005$ compared with previously published series, including 88% of patients undergoing unifocalization of major aortopulmonary collateral arteries (MAPCAs) and 12% of patients undergoing creation of an aortopulmonary (AP) window.

abnormal arborization of the pulmonary arteries or complete absence of the intrapericardial pulmonary arteries (encountered in 23% of the patients). The management of these patients has evolved to a midline unifocalization strategy incorporating as many of the MAPCAs as feasible into a central confluence. In this subgroup of patients, 64% are able to undergo a single-stage repair [3]. Ultimately, 90% of the patients have been able to undergo successful septation of the heart, with an RV/LV pressure ratio of 0.41 ± 0.12 immediately postoperatively and 0.36 ± 0.11 at 7 years. Actuarial survival at 5 years for this group was nearly 90%.

The results for patients undergoing creation of an aortopulmonary window can be contrasted with patients undergoing unifocalization as the primary method of treatment (Table 3). This comparison demonstrates that the rate of single-stage repair at the next operation after creation of the aortopulmonary window was only one-fifth (12% versus 64%; $p < 0.002$) that for patients undergoing unifocalization for MAPCAs. The rate of eventual achievement of successful septation (either single stage or multistage) was 55% versus 90%, respectively ($p < 0.004$). We interpret these results to suggest that hypoplastic pulmonary arteries and diminished pulmonary blood flow are markers for a less well developed pulmonary vascular bed, and we would hypothesize that the competitive flow associated with a "dual supply system" may have adverse effects on the development of the distal vascularity.

Rodefeld and associates [12] previously published an article from our surgical group summarizing the experience in the 1990s with the surgical creation of an aortopulmonary window. There was no operative mortality encountered in that series of 18 patients, as was seen in the current series as well. The early success of this approach may be related to the strict adherence of performing this procedure exclusively in patients with diminutive pulmonary arteries, thus circumventing the potential problems of pulmonary overcirculation or reperfusion lung injury, or both. The study by Rodefeld and associates documented a doubling of the diameter of the central pulmonary arteries during an average of 3.7 months after creation of the aortopulmonary window. This increase in pulmonary artery size documents the potential for growth of these vessels when supplied with an additional source of pulmonary blood flow.

One significant disparity between the data from Rodefeld and colleagues and the current study was the single-stage complete repair rate, which was 53% compared with 12%, respectively. The process that our group has used in selecting patients for the aortopulmonary window procedure has not changed during the interval of both studies. Specifically, the patients must have small (<2.5 mm) intrapericardial branch pulmonary arteries that are confluent, have normal arborization, and share dual supply to areas supplied by MAPCAs. However 1 aspect of our approach that we did change between studies was our standard for selecting which patients can proceed to complete repair based on the results of the intraoperative flow study. In the 1990s, we used a flow rate of 2.5 L/min/m² to the pulmonary arteries, accepting a mean pulmonary artery pressure less than 30 mm Hg. Our experience indicated that we were proceeding with VSD closure in some patients with higher pulmonary artery pressures by flow study who subsequently had unacceptably high right ventricle/left ventricle (RV/LV) pressures. As a consequence of this experience, we increased our threshold for VSD closure to a maximal flow rate of 3.0 L/min/m² and reduced the acceptable pulmonary artery pressure to 25 mm Hg. Rodefeld and colleagues documented a mean pulmonary artery pressure of 57 mm Hg at cardiac catheterization, which was similar to the mean of 53 mm Hg in the current study. These pulmonary artery pressures are essentially equivalent to systemic pressures, as expected with a direct communication between the aorta and pulmonary artery. Neither set of data was adjusted for pulmonic blood flow–systemic blood flow (Qp:Qs), which in theory could be calculated because the source of pulmonary blood flow for the aortopulmonary window and the MAPCAs is from the aorta. However in practice, the calculation of Qp:Qs would not be particularly meaningful because it would include both sources of pulmonary flow, and only flow from the aortopulmonary window will contribute to antegrade flow during an intraoperative flow study or after complete repair (in the absence of incorporation of MAPCAs during the repair). The relatively high mean pulmonary artery pressures would need to be associated with Qp:Qs values of approximately 2:1 in the earlier study and nearly 3:1 in the latter series to result in satisfactory values for the intraoperative flow study. It is likely that the major difference in single-stage complete repair rate observed between studies would be accounted for by the difference in intraoperative flow study criteria. It is also worth commenting that the ultimate success of any given strategy must be judged in the context of mid- to late-term outcomes and this will be strongly influenced by the relative pressure ratio of RV/LV pressure.

In a recently published series from the surgical group in Rome, Carotti and colleagues [8] found that the absence of confluent intrapericardial pulmonary arteries was associated with significantly better ratios of RV/LV pressure after complete repair (the mean RV/LV pressure was 0.48 ± 0.14, with univariate analysis demonstrating that patients with confluent intrapericardial pul-

monary arteries had worse hemodynamics). These results are consistent with the findings in our current study that the presence of confluent, intrapericardial pulmonary arteries was associated with a lower chance of complete repair. In contrast to these findings, Davies and associates [7] found that patients with confluent intra-pericardial pulmonary arteries had a higher chance of achieving complete repair. It is difficult to reconcile these contradictory results because they may reflect more of the institutional variations as opposed to an analysis of the underlying physiologic principles.

An alternative approach for the treatment of PA/VSD MAPCAs has been proposed by a group in Melbourne, Australia, which has advocated repair without incorpo-ration of MAPCAs [13, 14]. This approach is based on the fundamental premise that MAPCAs are dilatated bron-chial arteries that may have limited growth potential and preclude any long-term beneficial effects. As a conse-quence, this group has relied exclusively on the "Mel-bourne shunt" approach, and this is applied to all pa-tients who present with intrapericardial pulmonary arteries. The results of their "repair without unifocaliza-tion" strategy demonstrate that 58% of patients achieved complete repair. However the mean RV/LV pressure ratio was 0.62, meaning that nearly half of all patients had pressure values on the border of what would be consid-ered unacceptable hemodynamics (RV/LV ratio > 0.67). It is conceivable that the application of this approach to all patients results in pulmonary overcirculation, with the eventual development of pulmonary vascular obstructive disease in some patients.

In summary, the results of this study demonstrate that the early outcomes for the surgical creation of an aorto-pulmonary window in patients with small intrapericar-dial pulmonary arteries are excellent. However very few of these patients were amenable to complete repair at the second stage. Most of the patients required multiple reoperations to recruit sufficient pulmonary arborization to allow septation of the heart. We interpret these coun-terintuitive results to suggest that hypoplastic central pulmonary arteries and diminished pulmonary blood flow are markers for a less well developed pulmonary vascular bed.

Illustrations for this manuscript (Fig 2 through Fig 5) were created by Erin Anne Mainwaring.

References

1. Reddy VM, Liddicoat JR, Hanley FL. Midline one-stage complete unifocalization and repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. J Thorac Cardiovasc Surg 1995;109:832–45.
2. Reddy VM, McElhinney DB, Amin Z, et al. Early and intermediate outcomes after repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: experience with 85 patients. Circulation 2000;101:1826–32.
3. Malhotra SP, Hanley FL. Surgical management of pulmo-nary atresia with ventricular septal defect and major aorto-pulmonary collaterals: a protocol-based approach. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2009; 12:145–51.
4. Amark KM, Karamlou T, O'Carroll A, et al. Independent risk factors associated with mortality, reintervention, and achievement of complete repair in children with pulmonary atresia with ventricular septal defect. J Am Coll Cardiol 2006;47:1448–56.
5. Mumtaz MA, Rosenthal G, Qureshi A, et al. Melbourne shunt promotes growth of diminutive central pulmonary arteries in patients with pulmonary atresia, ventricular sep-tal defect, and systemic-to-pulmonary collateral arteries. Ann Thorac Surg 2008;85:2079–84.
6. Griselli M, McGuirk SP, Winslaw DS, et al. The influence of pulmonary artery morphology on the results of operations for major aortopulmonary collateral arteries and complex congenital heart defects. J Thorac Cardiovasc Surg 2004;127: 251–8.
7. Davies B, Mussa S, Davies P, et al. Unifocalization of major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect is essential to achieve excellent outcomes irrespective of native pulmonary artery morphol-ogy. J Thorac Cardiovasc Surg 2009;138:1269–75.
8. Carotti A, Albanese SB, Filleppelli S, et al. Determinants of outcome after surgical treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collat-eral arteries. J Thorac Cardiovasc Surg 2010;140:1092–103.
9. Ishibashi N, Shin'oka T, Ishiyama M, Sakamoto T, Kurosawa H. Clinical results of staged repair with complete unifocal-ization for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Eur J Cardio-thorac Surg 2007;32:202–8.
10. Bull K, Somerville J, Spiegelhalter D. Presentation and attrition in complex pulmonary atresia. J Am Coll Cardiol 1995;25:491–9.
11. Leonard H, Derrick G, O'Sullivan J, Wren C. Natural and unnatural history of pulmonary atresia. Heart 2000;84:499–503.
12. Rodefeld MD, Reddy VM, Thompson LD, et al. Surgical creation of aortopulmonary window in selected patients with pulmonary atresia with poorly developed aortopulmo-nary collaterals and hypoplastic pulmonary arteries. J Tho-rac Cardiovasc Surg 2002;123:1147–54.
13. Norgaard MA, Alphonso N, Cochrane AD, Menaham S, Brizard CP, d'Udekem Y. Major aorto-pulmonary collateral arteries of patients with pulmonary atresia and ventricular septal defect are dilated bronchial arteries. Eur J Cardiotho-rac Surg 2006;29:653–8.
14. d'Udekem Y, Alphonso N, Norgaard MA, et al. Pulmonary atresia with ventricular septal defects and major aortopul-monary collateral arteries: unifocalization brings no long-term benefits. J Thorac Cardiovasc Surg 2005;130:1496–502.

INVITED COMMENTARY

Mainwaring and colleagues [1] report on a challenging subgroup of patients with pulmonary atresia/ventricular septal defect and major aortopulmonary collaterals (ie, those having confluent diminutive intrapericardial branch pulmonary arteries, normal arborization, and dual pulmonary blood supply (12% of their series). When presenting with cyanosis, a sign of scanty collateral circu-lation, patients received construction of an aortopulmonary

---