Surgery for Congenital Heart Disease

Surgical creation of aortopulmonary window in selected patients with pulmonary atresia with poorly developed aortopulmonary collaterals and hypoplastic pulmonary arteries

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Objective: The morphologic characteristics of the pulmonary circulation vary widely in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals. Although we favor single-stage unifocalization and complete repair as the procedure of choice, a subgroup of patients who meet specific criteria have been treated with initial surgical creation of an aortopulmonary window.

Methods: Eighteen patients who were considered unsuitable candidates for single-stage repair underwent surgical creation of an aortopulmonary window. Selection criteria included the following: (1) presence of centrally confluent true pulmonary arteries 1.0 to 2.5 mm in diameter, with a well-developed peripheral arborization pattern; (2) multiple small aortopulmonary collateral vessels, most of which communicated with the true pulmonary arterial system; and (3) the presence of marked cyanosis.

Results: There were no early deaths, and the 2 late deaths were both unrelated to the procedure. Follow-up angiography (n = 17) demonstrated good growth of true pulmonary arteries in 15 of 17 cases (88%). Mean pulmonary artery diameter increased from 1.76 mm to 3.45 mm. Subsequent operations have been performed in 15 of 18 cases (83%). Intracardiac repair with or without additional unifocalization was achieved in 8 of these 15 (53%). Seven patients (47%) have undergone staged unifocalization as the next procedure; of these, 3 were able to have intracardiac repair. Thus 11 of the 15 patients who have undergone second operations (73%) have had complete intracardiac repair.

Conclusions: The initial surgical creation of an aortopulmonary window in carefully selected patients can increase the size of the true pulmonary arteries, making these patients better candidates for eventual intracardiac repair. The procedure should be avoided in patients with pulmonary overcirculation, a predominance of isolated supply collaterals, or true pulmonary arteries larger than 2.5 mm in diameter, and it is not applicable without a true pulmonary artery central confluence.

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The morphologic characteristics of the pulmonary circulation vary widely in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals. Although we favor single-stage unifocalization and complete repair as the procedure of choice, this approach demands that an adequate amount of native tissue be available for the required extensive reconstruction of the pulmonary arterial system. This native tissue can be derived from the pulmonary arterial system or the aortopulmonary collateral tissue. If adequate native tissue is not available from either or both sources, early single-stage complete repair is not appropriate. A subgroup of patients with inadequate native tissue for reconstruction who meet specific criteria have been treated with initial surgical creation of an aortopulmonary window by means of direct anastomosis between the aorta and diminutive main pulmonary artery trunk. These carefully selected patients have severely hypoplastic, centrally confluent true pulmonary arteries that, despite the central hypoplasia, have relatively well-developed peripheral pulmonary arterial arborization patterns. The collateral distribution in these cases is typically composed of multiple small vessels, most of which communicate with the true pulmonary arterial system. In this patient subgroup, an initial surgical aortopulmonary window was chosen as the best way to increase the size of the true pulmonary arteries, thereby increasing the amount of necessary native tissue and thus making these patients better candidates for eventual intracardiac repair. The selection criteria, surgical management, and subsequent surgical course of these patients were retrospectively reviewed.

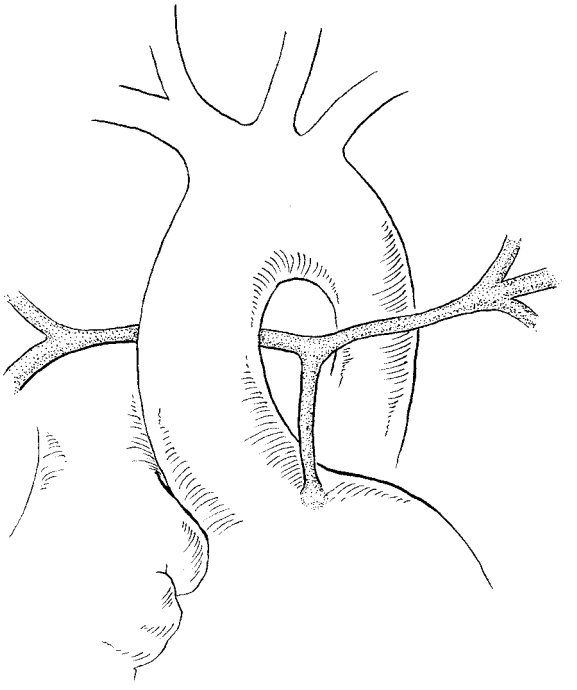

Figure 1. Pulmonary atresia with hypoplastic pulmonary arteries and aortopulmonary collaterals. Pulmonary arteries are frequently only 1 to 2 mm in diameter in this subgroup of patients. Dissection of pulmonary arteries to branch pulmonary artery bifurcation is critical to best position subsequent surgical aortopulmonary window.

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Methods

Selection Criteria and Patient Characteristics

Between August 1997 and October 2000, a total of 18 patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals who were considered unsuitable candidates for single-stage repair underwent initial surgical creation of an aortopulmonary window. The criteria for selection included the following: (1) presence of centrally confluent true pulmonary arteries 1.0 to 2.5 mm in diameter, with a well-developed peripheral arborization pattern; (2) multiple hypoplastic aortopulmonary collateral vessels, most of which communicated with the true pulmonary arterial system; and (3) the presence of marked cyanosis.

Age range at operation was 4 days to 8 months, with a mean age of 3.3 months. Mean number of major aortopulmonary collaterals was 3.75 (range 2-6). Associated anomalies included DiGeorge syndrome (n = 9), right-sided aortic arch (n = 3), double aortic arch (n = 1), complete atrioventricular canal (n = 1), Alagille syndrome (n = 1), and velocardiofacial syndrome (n = 1). The typical clinical picture was one of worsening cyanosis in a patient with unfavorable anatomy for single-stage repair. As is typical for these patients, systemic oxygen saturations varied. Most patients were maintained with supplemental oxygen support, and some received mechanical ventilatory support. Oxygen saturations before the operation ranged from 55% to 80%, with values typically in the 60% range. Several of these patients had been observed in early infancy for a planned unifocalization procedure, but because their conditions had become increasingly unstable they were selected for treatment with an aortopulmonary window.

Surgical Technique

Median sternotomy was used in all cases. After a limited pericardial incision, the great arteries were carefully dissected. The main pulmonary artery was carefully mobilized to its origin at the right ventricular infundibulum (Figure 1). The proximal left and right main pulmonary arteries were also carefully mobilized. Temporary neurovascular clips were placed on the branch pulmonary arteries, and the most proximal extent of the main pulmonary artery was divided as close as possible to its infundibular origin. If needed, the proximal margin could be oversewn or occluded with a surgical clip. The main pulmonary artery was then inspected, and the best position of the aorta was determined. Careful positioning was critical to avoid stretching and

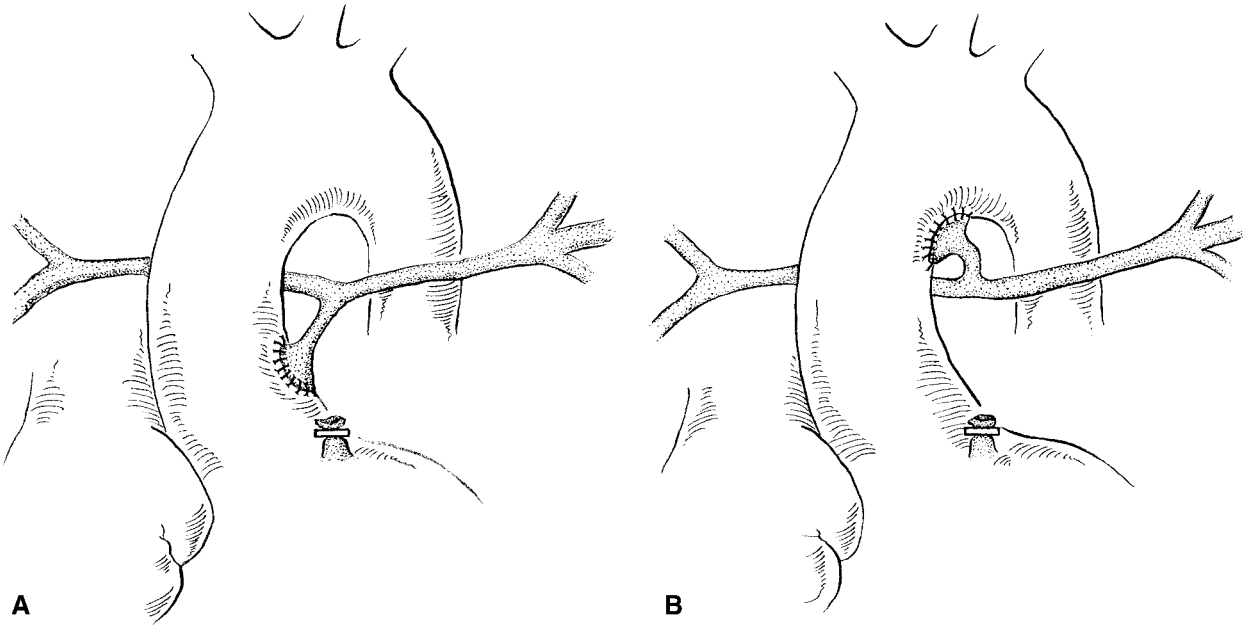

Figure 2. Surgical aortopulmonary window. Ideal location for anastomosis varies according to patient-specific anatomic variables and may be dictated not only by pulmonary artery anatomy but also by aortic size and position. A, Most common anastomotic position at left posterolateral aspect of ascending aorta, just above sinotubular junction. B, Less commonly, pulmonary artery confluence has better positional lie when located superiorly to undersurface of aortic arch.

distorting the right pulmonary artery branch where it passed behind the ascending aorta. Most commonly (n = 16/18, 89%) the pulmonary artery was anastomosed to the left posterolateral aspect of the ascending aorta just above the sinotubular junction, essentially adjacent to the original location of the main pulmonary artery (Figure 2). In occasional cases (n = 2/18, 11%), the pulmonary artery was better positioned by being anastomosed superiorly to the under side of the aortic arch. This decision may be heavily influenced by the anatomy of the ascending aorta and aortic arch, which can be quite variable in these patients. Careful and thoughtful selection of the anastomotic site cannot be overemphasized, because it is critical to avoid kinking or stenosis of the branch pulmonary arteries.

The goal of surgical aortopulmonary window was to create the largest anastomosis possible between the aorta and the diminutive main pulmonary artery trunk. The size was limited by the size of the proximal main pulmonary artery. The main pulmonary artery luminal diameter was typically consistent to the level of the infundibulum, and in some cases it widened considerably as a bulb at the pulmonary sinuses when a vestigial pulmonary valve was present. The main pulmonary artery was divided as proximally as possible to take advantage of this additional tissue when it was present. The proximal end of the divided main pulmonary artery was spatulated at its open end with a longitudinal incision approximately twice the diameter of the main pulmonary artery (Figure 3). The incision was made on the aspect of the pulmonary artery directly adjacent to the aorta. The patient received systemic heparinization (150 U/kg), and a side-biting clamp was applied to the anastomotic site. At completion of the anastomosis, the temporary neurovascular clips were removed from the pulmonary arteries and the partial-occluding aortic clamp was released. Diastolic blood pressure and systemic arterial saturations were carefully monitored. Typically, the diastolic blood pressure dropped by 5 to 10 mm Hg, and the systemic arterial oxygen saturations rose by 5% to 10%. The heparin was not reversed. After hemodynamic stability was ensured, the pericardium was loosely reapproximated and the sternotomy incision was closed. Cardiopulmonary bypass was not required in any case.

Results

All patients in this series underwent surgical creation of an aortopulmonary window as a primary operation, with the exception of 1 patient. In that case, a left lung unifocalization was performed to bring a stenotic at-risk collateral into the central system. This was followed by creation of an

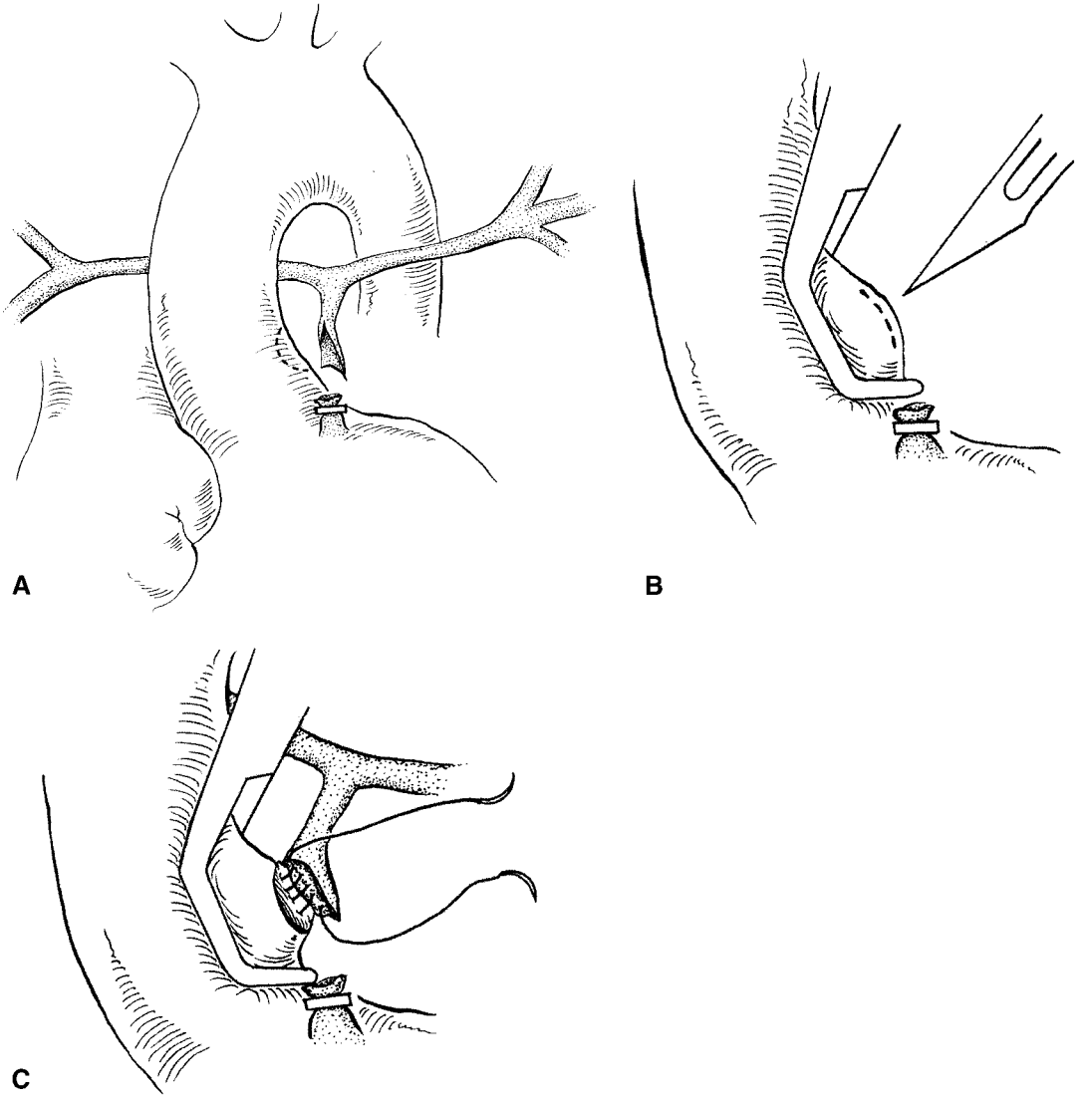

Figure 3. Surgical technique. A, The main pulmonary artery is divided as proximally as possible and spatulated with a longitudinal incision. B, It is important to carefully apply clamp such that sequestered portion of aorta is somewhat posterior to direct left lateral aspect of aortic circumference. C, Aortotomy is made, and anastomotic site is enlarged by excising aortic wall button. Anastomosis is performed with continuous 7-0 monofilament absorbable suture.

aortopulmonary window 3 weeks later. Seventeen patients underwent creation of an aortopulmonary window with native tissue-to-tissue anastomosis. In 1 unsuccessful case an aortopulmonary window was attempted, but a synthetic aorta-pulmonary artery shunt was required for technical reasons related to inadequate main pulmonary artery tissue for a satisfactory anastomosis. Because of the severely hypoplastic nature of the main pulmonary artery, this type of technical difficulty was fortunately rare. Revision or takedown of the window was not required in any case.

The postoperative courses of these patients were relatively uncomplicated, and postoperative management was routine. Inotropic requirements were minimal to none. The mean duration of ventilation was 3 days, with a range of 1 to 5 days. Fulminant pulmonary overcirculation or reperfusion injury was not observed in any case. In some instances, however, preventive measures were taken to manipulate pulmonary vascular resistance as part of a routine clinical management strategy in selected patients with evidence of mild pulmonary congestion on chest radiography or with increased pulmonary blood flow according to arterial blood gas analysis. Overt congestive heart failure, pulmonary

edema, or prolonged mechanical ventilation was not observed in the series.

There were no early or late deaths in the series directly related to creation of surgical aortopulmonary window. Late deaths unrelated to creation of aortopulmonary window occurred in 2 cases. One patient with Alagille syndrome, for whom the aortopulmonary window was viewed as a low-risk palliative procedure, was not a candidate for further surgical correction and died of hepatic complications approximately 4 months after the procedure. A second patient died of chronic pulmonary complications after completion repair. Morbidity was minor in the series. The most serious complication encountered was a transient neurologic event that occurred on the fifth postoperative day. Results of a neurologic workup were negative, and the patient was discharged 4 days later with no residual deficit. Although the cause of the event was unclear, it was not related to hemodynamic instability. Superficial wound infection occurred in 2 cases. In 1 case, postoperative hypoxemia was evaluated by early postoperative cardiac catheterization. This demonstrated a patent aortopulmonary anastomosis with flow in both branch pulmonary arteries. This patient, who had severe cyanosis for a prolonged period before being seen and had severely hypoplastic pulmonary arteries and collaterals, was the only patient in the series who had poor growth of the true pulmonary arteries after creation of an aortopulmonary window. In no case was thrombosis or branch pulmonary artery occlusion encountered.

Growth of the True Pulmonary Arteries

Patients were routinely reassessed by cardiac catheterization 2 to 4 months after creation of the aortopulmonary window. Follow-up angiography (n = 17) demonstrated good growth of the true pulmonary arteries in 15 of 17 cases (88%). Mean pulmonary artery diameter, as measured at the mid portion of the branch pulmonary arteries, increased from 1.76 mm before the operation to 3.45 mm after creation of the aortopulmonary window, with a mean interval to postoperative catheterization of 3.7 months (Figure 4). Mean branch pulmonary artery pressure was 57 mm Hg. Major stenosis of the proximal branch pulmonary arteries necessitating surgical reintervention was not encountered. In some cases, however, mild stenoses were dilated percutaneously. In 1 case in which no true pulmonary arteries were present, the aortopulmonary window was placed to a large bifurcating collateral vessel; growth of this collateral was moderate. In this instance, the patient had a preoperative diagnosis of centrally confluent true pulmonary arteries that was confounded by the large bifurcating collateral vessel. In 1 case, true pulmonary artery diameter did not increase. The reason for this is unclear. Cardiac catheterization demonstrated a patent anastomosis and flow in both pulmonary arteries. This patient ultimately required subsequent synthetic shunt placement to a centrally reconstructed pulmonary artery confluence.

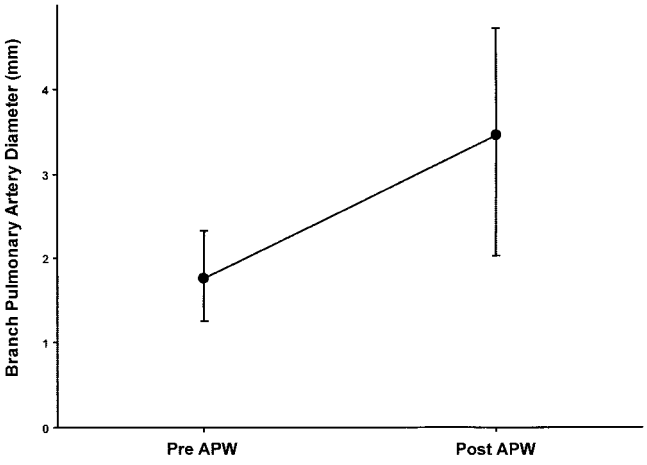

Figure 4. Preoperative versus postoperative branch pulmonary artery diameters, measured at midpoint in branch pulmonary artery. There were no differences between right and left branch pulmonary artery diameters either before or after creation of aortopulmonary window *(APW)*. *Points* represent mean; *error bars* indicate SD.

Status of Collaterals After Aortopulmonary Window

No change was observed in the collaterals that were isolated from the central pulmonary arterial system. In communicating collaterals with dual supply, change in collateral size is difficult to assess; however, dimensions appeared to be static in most cases. Coil embolization was used in collaterals with dual supply and competitive flow when there was no need to augment the caliber of the true pulmonary arterial system, or in collaterals that were remote from the surgical site and could not be surgically incorporated into the unifocalized neopulmonary artery. This typically included collaterals arising from the subclavian or intercostal arteries. Rates of coil embolization remained fairly low for communicating collaterals, because they were typically ligated at the time of surgical repair.

Nature and Timing of Subsequent Procedures

Subsequent operations have been performed in 15 of 18 patients (83%). Mean time to second operation was 4.6 months. Of the 3 patients who did not undergo subsequent operations, 1 has died of complications of Alagille syndrome and was not a candidate for additional procedures anyway, 1 is awaiting additional staged surgery, and 1 had transient neurologic complications and is unavailable for follow-up. Completion repair at the second operation was

achieved in 8 of 15 cases (53%) at a mean interval of 4.7 months. In 3 of these cases, collaterals could be simply ligated, and no unifocalization was required. Seven of these (n = 7/15, 47%) have had further staging; of these, 3 have had definitive repair. The remaining 4 patients remain in the process of staging toward complete repair. Thus the total number of patients in the series who have had definitive repair is 11 of 15 (73%).

Discussion

Although we favor single-stage unifocalization and complete repair as the procedure of choice, a subgroup of patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals who meet specific criteria have been treated with initial surgical creation of an aortopulmonary window. These patients were poor candidates for initial single-stage repair because of an inadequate amount of native tissue necessary to perform an adequate pulmonary arterial reconstruction, as determined by the presence of very small true pulmonary arteries in addition to multiple small major aortopulmonary collaterals.

Surgical creation of an aortopulmonary window is performed by direct anastomosis between the aorta and diminutive main pulmonary artery trunk. Although we have preferred to use the terminology "surgical aortopulmonary window," the procedure has been previously described as a "central end-to-side shunt."[1] This is the same technical procedure originally described by Watterson and colleagues[1] and advocated by that group as the procedure of choice for all patients with centrally confluent true pulmonary arteries. Although we agree that surgical creation of an aortopulmonary window has advantages with respect to synthetic shunts and right ventricular outflow tract patches[2-4] as a method of promoting true pulmonary arterial growth in this setting, we disagree with its routine use in all or most cases. We have developed stringent criteria for abandoning single-stage complete unifocalization and repair in favor of the aortopulmonary window, and these criteria were specifically chosen to avoid the unwanted consequences that can be associated with uncontrolled pulmonary blood flow. First, the patient must have profound cyanosis, defined as an arterial oxygen saturation of 82% or less. If the patient does not have such profound cyanosis, the native pathways to the pulmonary circulation are in aggregate adequate for normalization of the circulation, albeit with a major reconstructive effort. We believe that the more direct single-stage approach is of more benefit under these conditions. Second, the true pulmonary arteries must be severely hypoplastic. If they are larger than 2.5 mm in diameter, the unrestrictive aortopulmonary connection can cause overcirculation, with hemodynamic instability and pulmonary vascular injury. Third, the true pulmonary arteries must have a reasonably well-developed peripheral arborization pattern. If, on the other hand, the true pulmonary arteries arborize to an extremely limited degree, even with marked central pulmonary artery hypoplasia, the unrestrictive aortopulmonary connection can create distal pulmonary hypertension when the peripheral runoff is limited, theoretically damaging the pulmonary microvasculature. Fourth, the collateral system must be characterized by multiple small vessels, with most communicating with the true pulmonary arterial system. If the collaterals are large, single-stage repair is preferred. If numerous collaterals provide isolated blood supply to various lung segments, this implies that the true pulmonary arterial system has limited arborization. Under these conditions, an aortopulmonary connection creates the risk of pulmonary hypertension in the true pulmonary arterial system, and the isolated supply collaterals remain completely uncontrolled.

Pulmonary overcirculation was not encountered to any significant degree, in contrast to other reported series.[1] This is probably a function of our stringent selection criteria for this procedure, which avoided those patients who would be likely to have excessive pulmonary blood flow as a consequence of aortopulmonary shunting. Clinical reperfusion injury was not observed in any case, although in some instances routine preventive measures were taken to augment pulmonary vascular resistance as part of a routine clinical management strategy for those patients who had clinical evidence of pulmonary congestion on chest radiography or evidence of significant pulmonary blood flow according to arterial blood gas analysis.

Pulmonary arterial growth within 2 to 6 months helped make many of these patients more suitable candidates for subsequent single-stage or staged complete repair. Others have reported longer intervals to subsequent operation[1]; however, we maintained an aggressive early approach. This is in keeping with our philosophy of minimizing the duration of exposure of lung segments to systemic pressure. In most cases, pulmonary arterial growth was adequate for subsequent operation to be considered within a 3- to 6-month span. Patients underwent repeated angiography and were reassessed for subsequent repair according to our standard criteria.[5] Allowing a longer interval for assessment would have put these patients at substantial risk for development of vascular disease in the true pulmonary artery segments. Thus it is critical that the interval for reevaluation be limited, so that irreversible damage to the pulmonary microvasculature will not have occurred. Stenosis of the proximal right or left pulmonary arteries after creation of an aortopulmonary window was not seen in this series. This varies from other series, which report proximal stenosis rates exceeding 50%.[1] Adequate mobilization of the pulmonary arteries and placement of the window at an optimal

location on the aorta are essential to avoid proximal branch pulmonary artery stenosis.

In summary, the initial surgical creation of an aortopulmonary window in carefully selected patients can increase the size of the true pulmonary arteries and limit growth of small communicating major aortopulmonary collaterals, making these patients better candidates for eventual intracardiac repair. The procedure was associated with minimal morbidity, which in our opinion was directly related to the stringent selection criteria used in this series. The procedure should be avoided in patients with high-flow or isolated supply major aortopulmonary collaterals or with true pulmonary arteries larger than 2.5 mm in diameter, and it is not applicable without a true pulmonary artery central confluence.

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Discussion

Dr Carl L. Backer *(Chicago, Ill).* In light of the really excellent outcome with this strategy, which I think Mee has advocated for most patients, have you considered applying this more broadly to some of the patients for whom perhaps previously you would have advocated complete single-stage repair from the front, instead of staging them with this nice little interim operation?

Dr Rodefeld. The application of the procedure to a broader group of patients is not a good idea in our opinion. We strongly believe that the procedure should be limited to those patients who meet the selection criteria.

The reasons are as follows. If patients have true pulmonary arteries that are larger than 2.5 mm or their collaterals are larger, they likely are candidates for single-stage repair, and we will aggressively attempt to do a single-stage complete repair for such patients.

If patients have evidence of adequate pulmonary blood flow on preoperative evaluation or do not have severely hypoplastic pulmonary arteries, they would be at risk for pulmonary overcirculation. We have seen this in others' reports, and we believe that it is an important difference in our series.

Dr Marshall L. Jacobs *(Philadelphia, Pa).* I commend you on your superb results with these difficult cases. I have particular interest in the second part of your principal selection criteria, that is, the knowledge beforehand that this tiny pulmonary artery confluence has satisfactory arborization to many or most pulmonary segments. It may be that your imaging modalities are of superior quality, but I think that it is often the experience when this confluence of true pulmonary arteries is only visualized as a result of filling on an aortogram from some collaterals that one really does not know the arborization of the true pulmonary arteries until this procedure or an analogous central shunt procedure has been completed. You do your very early postoperative angiography, and only at that time do you learn which pulmonary segments are supplied by these true pulmonary arteries. Would you comment on whether that has been an observation among your patients? How does it bear on your selection criteria?

Dr Rodefeld. I agree that it can be difficult to judge the degree of arborization of the pulmonary arteries in these patients and that essentially cardiac catheterization, although as thorough as can be done, may not give 100% information. I think our approach is simply to get the best information possible.

The concern with limited arborization is that if these pulmonary arteries are brought into the systemic circulation that the lung segments supplied by those pulmonary arteries will be subjected to systemic flows. So we do try as best as possible to be certain that the arborization pattern is relatively full to both lungs.

Again, this can be difficult to completely determine, and I do not think that we have any specific way of determining that. We have had instances in which we have performed the operation and found that the anatomy was not completely as described, although I think that this is a problem encountered by most groups.

Dr John W. Brown *(Indianapolis, Ind).* This was a nice series, although we have approached this group of patients just slightly differently. We approach them from the front, but we use cryopreserved saphenous vein to do a central shunt. We think that saphenous vein is better suited for these low-flow states, and we have seen good growth. And we do not dissect out the pulmonary arteries at all. We just uncover that portion of the main pulmonary artery that we want to attach to the distal end of the shunt and leave everything else alone. Then, like you, we catheterize the patients 2 or 3 months after the operation to see what is going on. If they need unifocalization at that point, then that is what we do. If they do not, then we wait a little while longer before attempting a complete repair.

Dr Rodefeld. I think that there are a number of different ways to approach these patients. We consider the aortopulmonary window to be the best approach. The pulmonary arterial dissection I understand would be a concern, and we believe that it is important to dissect the pulmonary arteries completely simply because we are reanastomosing the main pulmonary artery to the aorta, and we want to be sure that there are no anatomic problems with the reconstruction.

Dr Christo I. Tchervenkov *(Montreal, Quebec, Canada).* I am particularly interested in the timing of the next operation. Since you have evidence that the creation of this aortopulmonary win-

dow results in growth of the native pulmonary arteries, what is the optimal time for the next intervention? Your median interval before the next procedure was 4.8 months. Why not wait somewhat longer, to get further growth of the native pulmonary arteries?

Dr Rodefeld. Essentially, our approach is that once the pulmonary arteries have grown to a sufficient size for us to proceed with additional repair or subsequent operation, we elect to go ahead and perform the repair. In other words, we do not want to theoretically subject the lungs to the systemic circulation for an additional period. So once the patient who has had an aortopulmonary window created has undergone catheterization, basically we reenter them into our standard selection criteria for additional unifocalization or repair.