SURGERY FOR CONGENITAL HEART DISEASE

MIDLINE ONE-STAGE COMPLETE UNIFOCALIZATION AND REPAIR OF PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND MAJOR AORTOPULMONARY COLLATERALS

Traditionally patients with pulmonary atresia, ventricular septal defect, diminutive or absent central pulmonary arteries, and multiple aortopulmonary collaterals have been managed by staged procedures necessitating multiple operations. We have taken a different approach to this lesion. Between August 1992 and March 1994, ten patients aged 1.43 months to 37.34 years (median 2.08 years) at the severe end of the morphologic spectrum of this lesion underwent a one-stage complete unifocalization and repair from a midline sternotomy approach. The median Nakata index of true pulmonary arteries was 50.0 (range 0 to 103.13) and they provided vascular supply to up to nine lung segments (median 5 segments). The number of collaterals per patient ranged from two to five with a median of four. The collaterals provided vascular supply to a median of 15 lung segments per patient (range 11 to 20). Complete unifocalization was achieved in all patients with emphasis on native tissue-to-tissue connections via anastomosis of collaterals to other collaterals and to the native pulmonary arteries. In only one patient (37.34 years old) was it necessary to use a non-native conduit for peripheral pulmonary artery reconstruction. The ventricular septal defect was left open in one patient (5 years old) because of diffuse distal hypoplasia and stenosis of the pulmonary arteries and the collaterals. The postrepair peak systolic right ventricular/left ventricular pressure ratio ranged from 0.31 to 0.58 (median 0.47). There were no early deaths. Complications were bleeding necessitating reexploration in one patient, phrenic nerve palsy in three patients, and severe bronchospasm in three patients. Follow-up (median 8 months, range 2 to 19 months) was complete in all patients. One patient was reoperated on for pseudoaneurysm of the central homograft conduit and then again for stenosis of the left lower lobe collateral. After this last operation at 13 months after the initial repair she died of a preventable cardiac arrest caused by pneumothorax. The patient with open ventricular septal defect underwent balloon dilation of the unifocalized pulmonary arteries, with a current pulmonary/systemic flow ratio of 1.4 to 1.8:1, and is awaiting ventricular septal defect closure. One other patient underwent balloon dilation of the reconstructed right pulmonary artery, with a good result. All survivors (9/10) are clinically doing well. This approach establishes normal cardiovascular physiology early in life, eliminates the need for multiple systemic–pulmonary artery shunts and use of prosthetic material, and minimizes the number of operations required. Long-term follow-up is essential to determine whether this approach will limit the need for further operations to central homograft conduit changes only. (J THORAC CARDIOVASC SURG 1995;109:832-45)

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Pulmonary atresia with ventricular septal defect (VSD) and major aortopulmonary (AP) collaterals is a rare and complex lesion in which great morphologic variability exists regarding the sources

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of pulmonary blood flow. The true central pulmonary arteries range from normal size to complete absence. Major AP collaterals, probably derived embryologically from the splanchnic vascular plexus,[1] are also highly variable in their size, number, course, origin, arborization, and histopathologic makeup.[2-6] A given segment of the lung may be supplied solely from the true pulmonary arteries, solely from the AP collaterals, or from both with connections between the two sources occurring at central or peripheral points and at single or multiple sites.[4, 6] In contrast, the intracardiac structure of this lesion is relatively straightforward, often with a single anteriorly malaligned VSD, well-developed right and left ventricles, and normal atrioventricular and ventriculoarterial connections.

The ultimate goal of surgical therapy in this lesion is to construct completely separated in-series pulmonary and systemic circulations. The prevailing management strategy for achieving this goal is to embark on a staged surgical reconstruction to centralize the multifocal pulmonary blood supply, recruiting as many lung segments as possible, then close the VSD and provide egress from the right ventricle to the "unifocalized" pulmonary arterial system.[7-10] This generally requires on average three separate operations.[7-9]

The most important physiologic factor signifying a favorable outcome for these patients after complete repair is the postrepair peak right ventricular pressure.[7, 11] This should be as low as possible. The peak right ventricular pressure depends greatly on the number of lung segments that are unifocalized and on the status of the pulmonary microvasculature in those segments. Another important factor is that the reconstruction must achieve unobstructed delivery of blood from the right ventricle to the pulmonary microvasculature. A number of impediments to achieving this ideal outcome exist. Lung segments can be lost for several reasons. The natural history of these major AP collaterals often follows a course of progressive stenosis and occlusion, sometimes making the segment of lung supplied by that collateral inaccessible at the time of unifocalization. Even if accessible, a long-standing severe stenosis of the collateral can lead to distal arterial hypoplasia and underdevelopment of preacinar and acinar vessels and alveoli.[4, 5] Also iatrogenic occlusion can occur when these collaterals are unifocalized in stages with nonviable conduits, sometimes resulting in loss of these segments. Finally major AP collaterals without obstruction can lead rapidly to pulmonary vascular obstructive disease in their supplied segments. Likewise, staged unifocalization necessitating the use of modified Blalock-Taussig or central shunts may result in pulmonary vascular obstructive disease.

Table I. *Patient demographics (n = 10)*

Parameter	*Value*
Age	1.43 mo-37.34 yr (median 2.08 yr; mean 5.96 yr; SD 11.36)
Sex, M:F	1:1
Weight (kg)	3.0-44.7 (median: 9.88; mean: 12.76; SD: 12.58)
BSA (m^2)	0.19-1.5 (median: 0.49; mean: 0.53; SD: 0.39)
Previous operations	2 patients*

SD, Standard deviation.

*A modified Blalock-Taussig shunt to the right upper lobe pulmonary artery was done in one and repair of cor triatriatum in the other patient. Both procedures were performed before referral to us.

It seems logical that the longer the microvasculature of a given lung segment is left to the hemodynamic vagaries of a major AP collateral, the more likely that pulmonary vascular obstructive disease will develop or involution will occur. Only the "perfectly stenosed" major AP collateral may allow normal distal development. Furthermore, the stenoses in the major AP collaterals are widely known to progress with time, suggesting that even a perfectly stenosed vessel is not likely to remain that way. Extending this logic, it seems clear that the sooner these hemodynamic vagaries can be removed the greater the likelihood that the largest number of healthy lung segments can be incorporated into the unifocalized pulmonary circuit. The pulmonary microvasculature taken in aggregate is healthiest at birth and declines thereafter.

One-stage complete unifocalization and repair early in life gives the greatest chance of achieving a healthy and complete pulmonary vascular bed. In this report we review our experience with this approach in the first 10 patients.

Methods

Patients. From August 1992 to March 1994, all patients referred to us with a diagnosis of pulmonary atresia VSD, and major AP collaterals were managed with the present approach. The demographic characteristics of these 10 patients are summarized in Table I. Their ages varied widely but five (50%) were less than 9 months old. Four patients were between 3 and 9 years old and there was one 37.34-year-old patient. The diagnosis was established by cardiac catheterization and angiocardiography. Whenever possible, all collaterals were identified by selective angiog-

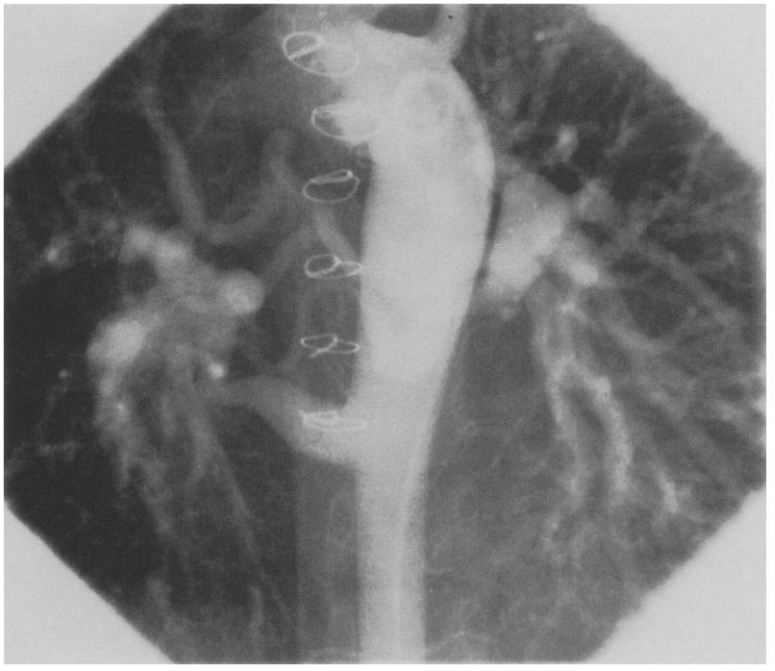

Fig. 1. Angiogram of a 3.5-year-old patient showing multiple major AP collaterals arising from the descending aorta. True pulmonary arteries were absent.

Table II. *Characteristics of MAPCAS (n = 10)*

Parameter	*Value*
Number of MAPCAS per patient	2-5 (median 4; mean 3.75; SD 0.95)
Number of segments supplied by MAPCAS	11-20 (median 15; mean 15.5; SD 3.47)
Number of MAPCAS unifocalized	2-5 (median 4; mean 3.75; SD 0.95)
Mean pressure in the collaterals (n = 15)	7-88 mm Hg (median 16 mm Hg)

MAPCAS, Major AP collaterals; *SD,* standard deviation.

Table III. *PA morphology (n = 10)*

Parameter	*Value*
RPA size (mm)	0-8.35 (median 2.93; mean 3.19; SD 2.50)
LPA size (mm)	0-6.26 (median 2.79; mean 3.03; SD 2.22)
Nakata Index	0-103.13 (median 50.0; mean 49.69; SD 42.33)
Number of segments supplied by PAs	0-9.0 (median 5.0; mean 4.5; SD 3.47)

PA, Pulmonary artery; *RPA,* right pulmonary artery; *LPA,* left pulmonary artery; *SD,* standard deviation.

raphy, and pressure measurements were made. An effort was made in all patients to identify the native pulmonary arterial anatomy and the hemodynamics and morphologic characteristics of the major AP collaterals. All patients had preoperative echocardiography. The operation was electively scheduled shortly after presentation and workup. If the diagnosis was made in the newborn period, the patient was discharged shortly after birth and the repair was done electively in the first 3 to 6 months of life. Older patients, before referral to us, had been followed up for variable periods because they were thought to have either unrepairable or well-balanced physiology.

The morphologic characteristics of the collaterals and the true pulmonary arteries are given in Tables II and III, respectively. These characteristics justify placing these patients in the severe end of the morphologic spectrum of pulmonary atresia, VSD, and major AP collaterals (Fig. 1). In two (20%) patients the true pulmonary arteries were completely absent and in one 37.34-year-old patient the pulmonary arteries were stringlike with a Nakata index[12] of 3.34. Pressure measurements were made in the individual collaterals by selective catheterization (Table II). This was not possible in some very young infants and was not attempted when the collateral arteries were significantly stenotic at their origin. All patients had a single VSD, of the malalignment type in nine patients and of the subarterial type in one patient. The atrial septum was intact in five of the 10 patients and an atrial septal defect (4/10) or patent foramen ovale (1/10) was present in the rest. The surgical procedures performed and the cardiopulmonary bypass data are given in Table IV.

Technique. Through a generous midline incision, a median sternotomy and a subtotal thymectomy were performed. The right pleura was widely opened anterior to the phrenic nerve and the right lung was lifted out of

Table IV. *Operative data (n = 10)*

Parameter	*Value*
Complete unifocalization	10 patients
VSD closure	9 patients
RVOT conduit (valved allograft)	10 patients
Create PFO*	4 patients
ASD closure/create PFO	5 patients
Cardiopulmonary bypass time (min)†	mean 250.8; SD 50.24
Aortic crossclamp time (min)	mean 80.11; SD 25.97

VSD, Ventricular septal defect; *RVOT,* right ventricular outflow tract; *PFO,* patent foramen ovale; *ASD,* atrial septal defect; *SD,* standard deviation.

*These patients had intact atrial septum.

†Most of the time on bypass the patient was at normothermia with the heart beating.

the pleural cavity. The descending aorta was exposed in the posterior mediastinum and all the collaterals from it were identified and dissected. Similarly, the left pleura was opened and the left-sided collaterals were identified and dissected. Avenues for collateral rerouting were developed by opening the pleura on both sides posterior to the phrenic nerves in the hilar regions. After this, the pericardium was opened and a large piece was harvested and fixed in glutaraldehyde. Attention was then directed to the central mediastinum, and the native pulmonary arteries, if present, were dissected out. Any further collaterals from the upper descending aorta were identified and dissected in the "subcarinal" space (between the tracheobronchial angle and the roof of the left atrium) by an approach between the right superior vena cava and the aorta (Fig. 2). The floor of the pericardial reflection in the transverse sinus was opened and the posterior mediastinal soft tissues were dissected to expose the aortic segment and the collaterals in this region. This was an important maneuver for gaining access to collaterals, which typically can arise from this location. Opening this space also provided an avenue for collateral rerouting for direct tissue-to-tissue anastomosis during unifocalization, which otherwise would have been impossible. Additionally, in some cases collaterals arising from the aortic arch or the neck vessels were exposed and dissected. All collaterals were snared to achieve control before cardiopulmonary bypass was begun.

As many collaterals as possible were permanently ligated at their origin, mobilized, and unifocalized without cardiopulmonary bypass. When the patient's oxygenation reached a compromising level, cardiopulmonary bypass was instituted and the rest of the collaterals were unifocalized, at normothermia with the heart beating. A warm calcium-supplemented blood prime was used in the pump circuit to maintain normal cardiac function. During the unifocalization process the emphasis was on avoiding synthetic or allograft conduits in the periphery and on achieving unifocalization by native tissue-to-tissue anastomosis. One or more of the following techniques of unifocalization were generally used in these patients:

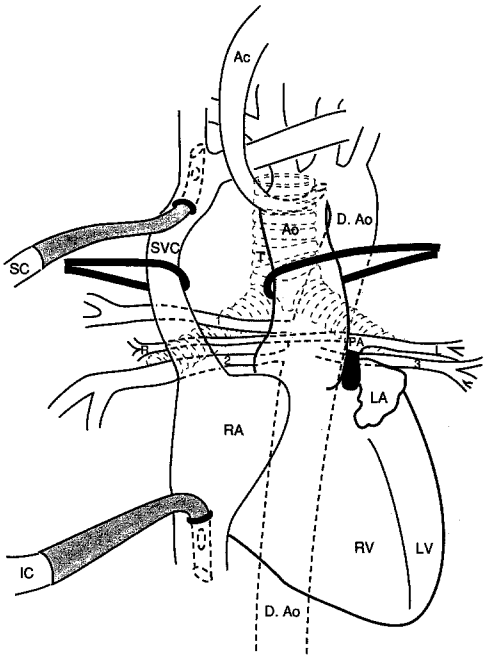

Fig. 2. Diagram of the transverse sinus approach for the dissection, rerouting, and unifocalization of the major AP collaterals. *Ac,* Aortic cannula; *Ao,* ascending aorta; *D.Ao,* descending aorta; *IC,* venous cannula in the inferior vena cava; *L,* true left pulmonary artery; *LA,* left atrium; *LV,* left ventricle; *PA,* long-segment pulmonary atresia; *R,* true right pulmonary artery; *RA,* right atrium; *RV,* right ventricle; *SC,* venous cannula in the superior vena cava (*SVC*); *T,* trachea; *1, 2, and 3,* major AP collaterals. In this diagrammatic representation the major AP collaterals are all shown anterior to the tracheobronchial tree. However, these collaterals often have variable relation to the tracheobronchial tree and the esophagus. Therefore unifocalization should be individualized to achieve the best lie of the major AP collateral.

1. Side-to-side anastomosis of the collateral to the central pulmonary arteries thereby augmenting the hypoplastic central pulmonary arteries (Fig. 3, *A* to *C*)
2. Side-to-side anastomosis of collateral or collateral to peripheral native pulmonary artery
3. End-to-side anastomosis of collateral to collateral or collateral to native pulmonary artery (Fig. 3, *B* and *C*)
4. Anastomosis of button of aorta (giving rise to multiple unobstructed collaterals) to the native pulmonary arteries. (Fig. 4, *A* and *B*)
5. End-to-end or end-to-side anastomosis of collateral to central conduit (Fig. 5, *A* and *B*)

These anastomoses were achieved directly by bringing collaterals above or below the lung hilum or through the transverse sinus, using as much of the collateral length as possible. Collateral length was given the highest priority to achieve tissue-to-tissue anastomosis. For example, if a

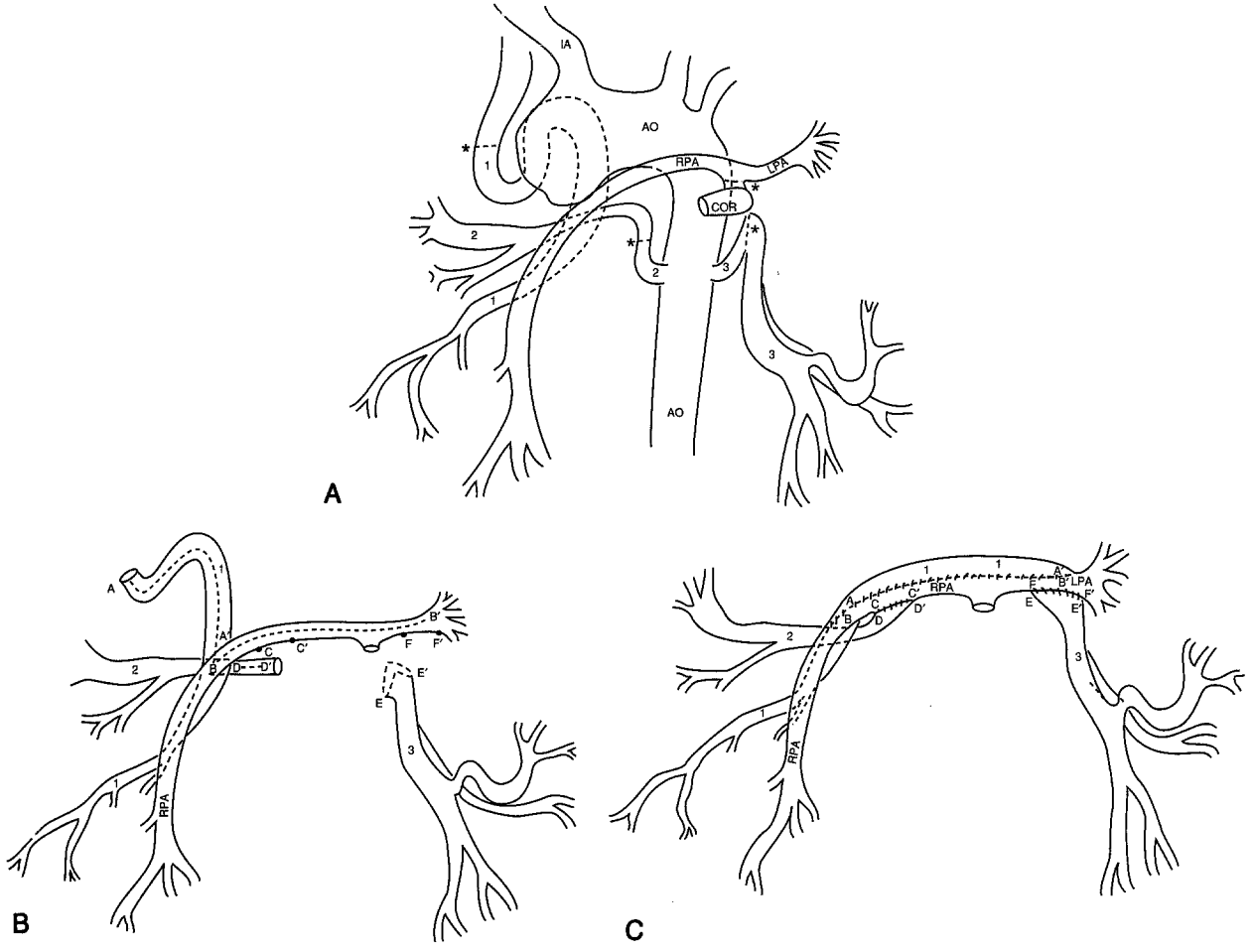

Fig. 3. Diagrams of the anatomy of the major AP collaterals and true pulmonary arteries and the techniques of unifocalization in a 3.5-month-old infant weighing 3.5 kg. **A,** Schematic representation of the major AP collaterals and true Pulmonary Arteries. *AO,* Aorta; *COR,* coronary collateral; *IA,* innominate artery; *LPA,* left pulmonary artery; *RPA,* right pulmonary artery, *1,* large and tortuous major AP collateral arising from the carotid artery; *2 and 3,* major AP collaterals arising from the descending aorta. *Site of ligation and division of the major AP collaterals. **B,** *A-A', D-D', E-E',* the major AP collaterals are filleted open along the broken lines; *B-B',* the true right and left pulmonary arteries are filleted open from hilum to hilum. **C,** *A-A' to B-B',* major AP collateral 1 is filleted open along its length all the way to the hilum and is anastamosed to the true pulmonary arteries, thereby augmenting the pulmonary arteries from hilum to hilum; *C-C' to D-D' and F-F' to E-E',* major AP collaterals 2 and 3 are anastomosed to the augmented true pulmonary arteries in an end-to-side fashion. Unifocalization and central pulmonary artery reconstruction are thus achieved without the need for non-native material.

discrete stenosis was present in the midportion of a collateral, the entire collateral would still be used. The stenosis was managed by side-to-side reconstruction at the necessary level or, if that was not possible, by patching. Even collaterals that had dual supply to a lung segment along with true pulmonary artery supply were unifocalized to build up the size of the reconstructed pulmonary arteries. The important concepts necessary to achieve this type of unifocalization are flexibility regarding reconstruction, aggressive mobilization, maximizing length of the major AP collaterals, and creative rerouting.

After complete unifocalization was achieved the patient was cooled to moderate hypothermia (about 25° C). The aorta was then cross-clamped and cardioplegic solution was given. However, occasionally, particularly difficult aspects of unifocalization were completed with the aid of hypothermia. A longitudinal ventriculotomy was then made in the right ventricular infundibulum and the hypertrophied muscle bundles were resected. The VSD was closed with a glutaraldehyde-fixed autologous pericardial patch or a Dacron patch with interrupted mattress pledget-supported Ti-Cron sutures (Davis & Geck, Danbury, Conn.).

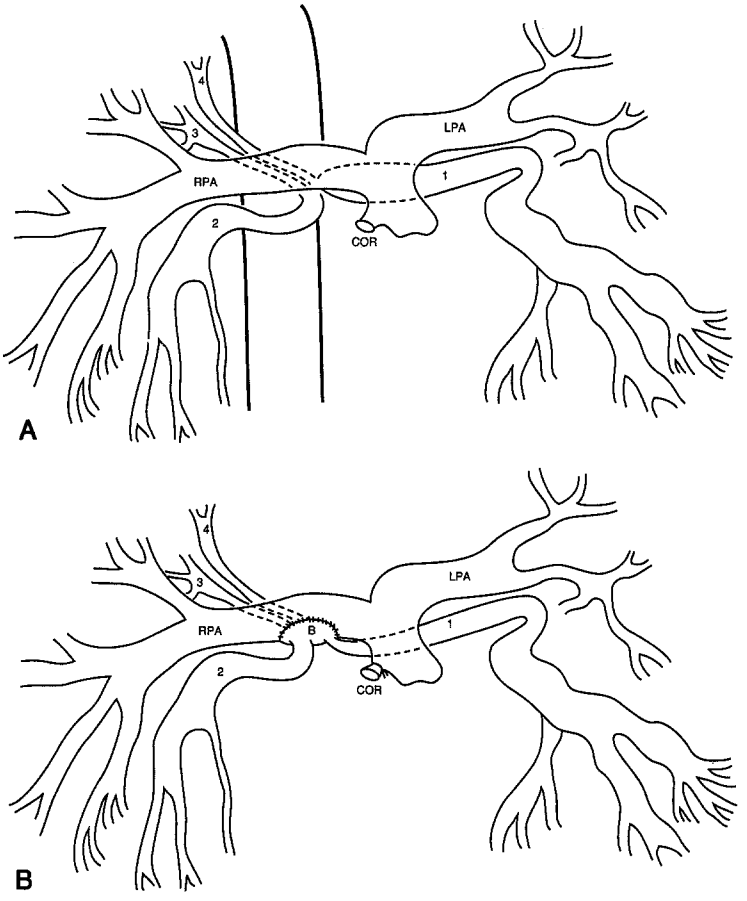

Fig. 4. Diagrams of the anatomy of the major AP collaterals and true pulmonary arteries and the technique of unifocalization in a 9.2-month-old infant. **A,** Schematic representation of the anatomy of major AP collaterals and true pulmonary arteries. *COR,* coronary collateral; *LPA,* left pulmonary artery; *RPA,* right pulmonary artery; *1, 2, 3 and 4,* major AP collaterals. **B,** *B,* Button of aortic wall with all the major AP collaterals is anastomosed to the right pulmonary artery, thereby achieving complete unifocalization.

The right atrium was opened to inspect the atrial septum. An atrial septal defect or patent foramen ovale, if present, was partially closed to leave a small unidirectional interatrial communication as a "popoff" valve for venous blood in case of postoperative right ventricular dysfunction. In some cases with intact atrial septum a small one-way interatrial communication was created. At this stage rewarming was started.

An allograft valved conduit was tailored and used in all cases to connect the right ventricle to the reconstructed neo-pulmonary arterial system. The distal conduit was anastomosed to the reconstructed pulmonary arteries. If needed, a distal tongue of tissue was shaped to augment the reconstructed central branch pulmonary arteries. In a total of 40 unifocalized collaterals, only one was reconstructed with a nonviable conduit (expanded polytetrafluoroethylene). This collateral supplied three lung segments in a 37.34-year-old patient. In two of the three patients with absent or stringlike true pulmonary arteries a second nonvalved allograft conduit was necessary to reconstruct the central left and right pulmonary arteries. One of these patients was the 37.34-year-old patient. In the second patient, in whom growth potential was an issue, the hilar regions were reconstructed with only natural tissue by means of the techniques described, and this second conduit served as the main left and right pulmonary arteries only, with the conduit limited to the pericardial cavity. In the third patient collaterals alone were used to reconstruct the central main, right, and left pulmonary arteries, without the need for a second conduit. The proximal right ventricle–to–conduit anastomosis was performed with a running Prolene suture (Ethicon, Inc., Somerville, N.J.). A pressure-monitoring catheter was placed through the right ventricular free wall into the pulmonary arteries across the right ventricular outflow tract. The right ventriculotomy was then closed with a pericardial or an allograft patch shaped like a hood extending from the proximal conduit onto the right ventricle.

After separation from cardiopulmonary bypass, aortic, right ventricular, pulmonary arterial, and atrial pressures were measured continuously. A transesophageal echocar-

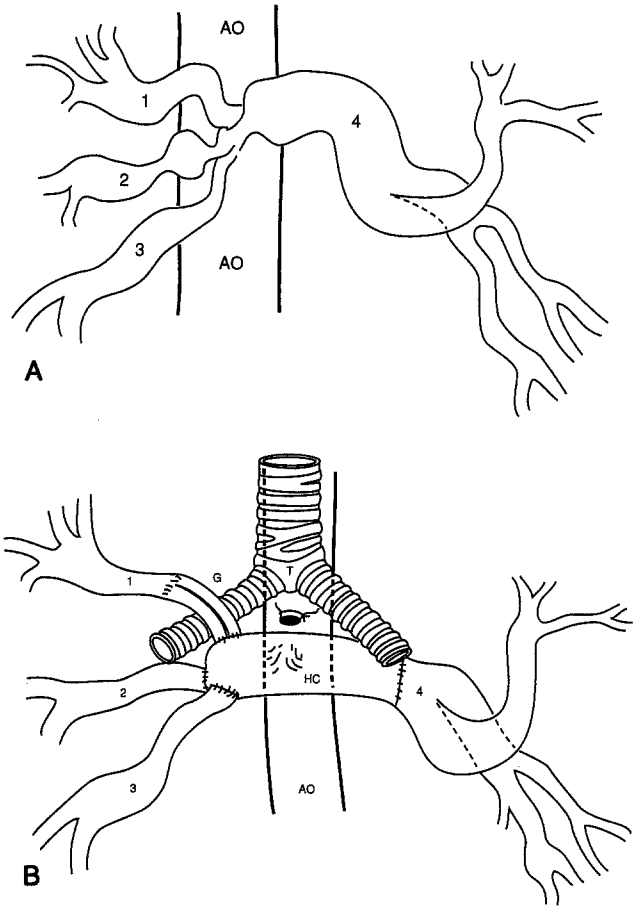

Fig. 5. Schematic representations of the anatomy of the major AP collaterals and the technique of unifocalization. **A,** *1, 2, 3, and 4,* major AP collaterals arising from a short segment of the descending aorta are stenotic at their origins. *AO,* Aorta. **B,** Each of the major AP collaterals *(1, 2, 3, and 4)* is anastomosed to an allograft conduit *(HC)*, which is now the central pulmonary artery. A 6 mm ePTFE tube graft *(G)* is interposed to anastomose collateral *1* (which was posterior to the right bronchus is translocated anteriorly) to the allograft conduit. Non-native material was used in this 37.34-year-old patient, in whom growth was not an issue. *T,* Trachea.

diogram was performed to ensure that there were no significant residual defects. Superior vena caval and pulmonary arterial oxygen saturations were estimated to rule out residual VSD.

Bilateral pleural and mediastinal tube drains were placed and the sternum was closed. In one infant the sternum was electively left open because of edema. The sternum was closed uneventfully on the second postoperative day.

Follow-up. At discharge all patients had an echocardiogram to assess right ventricular performance and the overall reconstruction. All patients have been followed up by clinical examination, and an echocardiogram or a catheterization was done at the discretion of the referring cardiologist. Follow-up evaluation was achieved in all patients by primary physician contact or by direct patient contact. Follow-up ranged from 2 months to 19 months with a median of 8 months. Five patients had a cardiac catheterization at a median follow-up of 7 months.

The data are expressed as median, mean, standard deviation, and range. Student's paired *t* test was performed with the SPSS statistical package (SPSS, Inc., Chicago, Ill.).

Results

Early results. All 10 patients survived the initial operation. In one patient, a 5-year-old child who had been profoundly cyanotic for several years, the VSD was not closed because both the native pulmonary arteries and the collaterals beyond the stenotic areas were diminutive and it was deemed during the operation that the risk of unacceptable postrepair

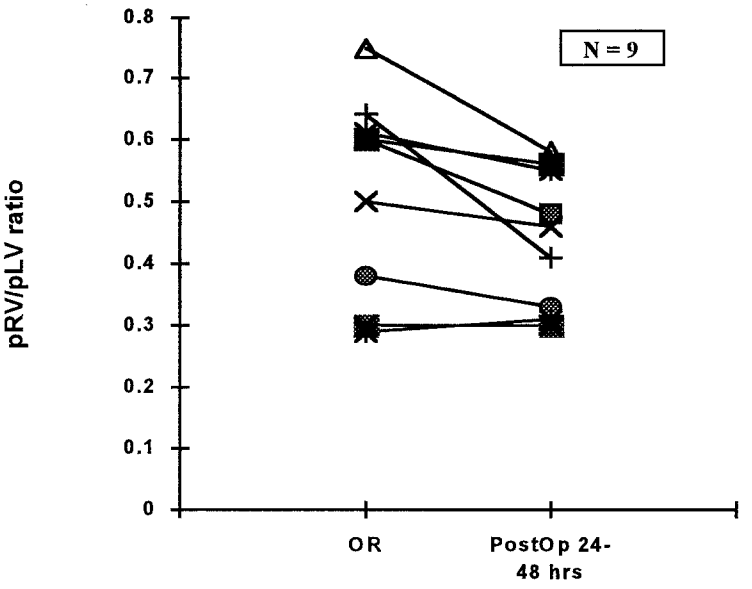

Fig. 6. The trend of the pRV/pLV. Mean pRV/pLV 24 to 48 hours after repair was significantly ($p < 0.02$) lower than that measured in the operating room.

peak right ventricular pressure would be high. This patient required reexploration for postoperative bleeding.

The ratio of peak systolic pressure in the right ventricle to the peak systolic pressure in the left ventricle (pRV/pLV) in the operating room ranged from 0.29 to 0.75 (mean 0.52; standard deviation 0.16; median 0.6). At 24 to 48 hours after the operation the pRV/pLV was 0.31 to 0.58 (mean 0.46; standard deviation 0.10; median 0.47). Postoperative pRV/pLV tended to decrease significantly ($p < 0.02$) from that measured in the operating room (Fig. 6). The postoperative right ventricular pressures were obtained during withdrawal of the surgically placed pulmonary arterial catheter. No significant right ventricle–pulmonary artery conduit gradient was detected in any of these patients.

Minimal inotropic support (dopamine ≤ 5 μg/kg per minute) was required for a median duration of 2 postoperative days (range 1 to 6 days). In only one patient moderate doses of two inotropic agents were required. This patient was also very edematous at the end of the bypass run and the chest was left open. On the second postoperative day the chest was closed uneventfully and this did not contribute to any increased postoperative morbidity.

Postoperative mechanical ventilatory support was required for a median duration of 2.21 days (range 0.4 to 25 days). In four of the 10 patients (all younger than 9 months), mechanical ventilation for more than 10 days was required. The causative factors were diaphragmatic paralysis (3/4) and frequent episodes of bronchospasm (3/4). Diaphragmatic plication before extubation was required in two patients. Examination with a bronchoscope revealed diffuse inflammation of the tracheobronchial tree in one patient who had episodes of severe bronchospasm.

The median stay in the intensive care unit was 2.21 days and the range was 1.5 to 36 days. All patients were discharged from the hospital 5 to 40 days after the operation. The median hospital stay was 14.5 days.

A discharge echocardiogram was available in all 10 patients. All patients had good right ventricular function. Tricuspid regurgitation was absent in five patients, trivial to mild in four patients, and moderate in one patient. Conduit insufficiency was mild in five patients, trivial in two patients, and absent in three patients. There were no residual intracardiac defects. At discharge the arterial oxygen saturation of patients in whom total repair was achieved ranged from 93% to 100% (median 95%). Only one patient was discharged with home oxygen, owing to pulmonary problems related to prematurity.

Follow-up. All living patients were assessed in March or April 1994. Follow-up ranged from 2 to 19 months with the median follow-up duration being 8 months. Reinterventions were required in two of nine (22.2%) patients who had complete repair. In

one patient (the first in our series), 4 months after repair a large pseudoaneurysm resulting in severe right ventricular failure developed at the proximal conduit anastomosis. The conduit was successfully replaced. The pRV/pLV at this time was 0.6. A month later this patient was evaluated by cardiac catheterization and a balloon angioplasty of the right pulmonary artery was performed with a satisfactory result. Follow-up lung scan revealed poor perfusion of the left lower lobe. This was consistent with the nonvisualization of the left lower lobe collateral, which had been anastomosed to the right lower lobe collateral across the midline. The left lower lobe collateral was successfully reimplanted into the left pulmonary artery system via a left thoracotomy approach. During an otherwise uneventful recovery in the postoperative period, the patient had a cardiopulmonary arrest as a result of an undiagnosed pneumothorax. Despite successful resuscitation from the arrest the patient eventually died of multiorgan failure 13 months after the original operation. This was the only death in this series. In the second patient, 4 months after the operation a follow-up echocardiogram suggested accelerated flow in the right pulmonary artery system, moderate conduit insufficiency, mild right ventricular enlargement, trivial tricuspid regurgitation, and a predicted right ventricular pressure of 46 mm Hg. Cardiac catheterization and angiography were performed to evaluate the right pulmonary artery. The angiogram showed an unobstructed left pulmonary artery system and long-segment stenosis of the unifocalized right upper lobe collateral. A successful dilation of this vessel was performed with an increase in size of the artery from 2 to 4 mm with excellent flow characteristics. Four months later this patient underwent another catheterization because of decompensation during a severe respiratory syncytial viral pneumonia. No significant pulmonary arterial stenoses were detected. The pRV/pLV was 0.51. Cardiac function recovered fully after the respiratory syncytial viral pneumonia resolved.

Two other patients who had had complete repair underwent cardiac catheterization and angiography 3 weeks after the operation because of inability to be weaned from ventilatory support. Both these patients subsequently underwent diaphragmatic plication. In one patient aged 1.4 months there were no residual defects and the pRV/pLV was 0.33 (postoperative pRV/pLV = 0.31). In the other patient aged 3.5 months the angiogram revealed hypoplastic areas in the collaterals used to reconstruct the distal right and left pulmonary arteries. The pRV/pLV at this time was 0.63. Because it was only the third postoperative week we elected not to perform an angioplasty. A subsequent catheterization 2 months later showed significant growth of the narrowed areas and angioplasty was deemed unnecessary (pRV/pLV = 0.6). This decision was substantiated by perfusion lung scan findings. Two weeks after the operation there had been 75% perfusion to the right lung and 25% to the left lung. A follow-up scan at the time of the most recent catheterization revealed a right/left lung perfusion ratio of 58%:42%.

The only patient in whom the VSD was not closed underwent planned angioplasty of both the right and left unifocalized pulmonary arterial systems on three occasions. The most recent catheterization data on this patient revealed a pulmonary/systemic flow ratio of 1.4 to 1.8:1.0. One more session of angioplasty followed by possible VSD closure is planned for her.

The last follow-up echocardiogram (at a median interval of 1 month after the operation) in all patients did not show any significant change from the discharge echocardiogram. Of the nine survivors, all are clinically doing well without heart failure. At most recent follow-up only three patients are receiving cardiac medication in the form of digoxin and frusemide (Lasix). One of these is the patient with the open VSD. The second patient is a premature infant with velocardiofacial syndrome and lung disease who had weighed 1.6 kg at birth. The third patient was without cardiac medication for about 5 months after repair until he had a severe respiratory decompensation as a result of respiratory syncytial viral pneumonia.

Discussion

Pulmonary atresia with VSD and major AP collaterals is an uncommon lesion comprising 25% of all cases of pulmonary atresia and VSD.[6] The hallmark of this lesion is lack of adequate central and peripheral pulmonary arteries with the majority of the pulmonary parenchyma being supplied by major AP collaterals. This multifocal nature of the pulmonary vascular supply has complicated the management of this lesion, which otherwise has relatively straightforward intracardiac morphology. Before the systematic hemodynamic and morphologic studies of the collateral supply by Macartney and associates,[13] Haworth,[14] Haworth and Macartney,[4] Haworth and colleagues,[15] Thiene and coworkers,[16] and Rabinovitch and associates,[5] most of these

patients were either managed medically or treated by permanent palliation. The concept of unifocalization of the multiple sources of vascular supply to the lungs was suggested by Haworth and Macartney.[4]

Initial successful case reports[17-19] of surgical management of pulmonary atresia, VSD, and major AP collaterals were followed by the reports of larger experiences.[7, 8, 9, 20] Over a 5-year period Puga and coworkers[7] managed 38 patients by a staged approach. Not including intracardiac repair, on average 1.42 procedures were required per patient to unifocalize the major AP collaterals, with a 7.9% mortality. At the end of 5 years 23 patients (60%) had undergone a total repair with two deaths and three patients had a poor hemodynamic result with a pRV/pLV of more than 0.85. Four other patients were considered to have unrepairable anomalies because of unsuitable pulmonary arteries or major AP collaterals. Iyer and Mee,[8] in their experience with 58 patients over a 10-year period, performed 2.1 staging procedures (excluding repair) per patient to complete the unifocalization, with a mortality of 10.3%. Total repair was possible in only about 52% patients. At the end of their study period 22 of 58 (38%) patients were either considered to have anomalies unsuitable for repair ($n = 12$) or they had died ($n = 10$). Similarly, using a staged approach in 34 patients, Sawatari and associates[9] at the end 6 years had performed complete repair in 16 patients (47%). Of those 16 patients, two died and four had an unacceptable pRV/pLV. Overall only 30% of their patients had a good result at the end of 6 years. Only 12% (3/26) of patients reported on by Sullivan and coworkers[10] underwent a definitive operation over a 7-year period. Although these reports represent groundbreaking landmarks in the management of pulmonary atresia, VSD, and major AP collaterals, the substantial cumulative mortality and frequent failure to achieve total repair emphasizes the shortcomings of a staged approach. Additionally, such an approach also does not take into account the substantial number of patients who either died before a first procedure or were ruled unsuitable to enter into a staged program. Taking all of these points of attrition into account, it appears that only 20% to 30% of a cohort of newborn infants with this combination of anomalies will have complete repair with acceptable right ventricular hemodynamics if a delayed staged approach is taken.

Failure to recruit enough lung segments into a unifocalized pulmonary vascular supply to assure acceptable postrepair hemodynamics can be due to several reasons. The longer the collaterals are exposed to systemic pressure and flow, whether by their natural connections or by systemic arterial shunts, the more likely it is that myointimal hyperplasia will develop and lead to eventual occlusion of these collaterals. Lung segments distal to this may become inaccessible. Another mechanism of loss of lung segments is by the development of pulmonary vascular obstructive disease in the segments supplied either by unobstructed collaterals or by large systemic arterial shunts commonly performed during various staging procedures. Finally, scarring and distortion of the pulmonary arteries from multiple operations and synthetic conduit usage in the peripheral pulmonary arterial system may result in occlusion and further loss of recruitable lung systems. Early one-stage complete repair favorably affects all of these mechanisms.

It has been recommended that the minimum requirements for a single-stage complete surgical repair include a central pulmonary arterial area at least 50% of normal and pulmonary arteries connected to at least one whole lung worth of pulmonary segments.[7] In all patients who do not meet these criteria, a staged approach was strongly recommended.[7] Also complete repair in infancy has been discouraged as a general principle. Our experience does not support these contentions. A single-stage complete unifocalization was achievable in 100% of our patients and single-stage unifocalization with complete intracardiac repair was achievable in 90% regardless of the true pulmonary arterial anatomy. None of our patients had a true pulmonary arterial supply to more than nine segments, and on an average only five segments were supplied (see Table III). Furthermore, the median Nakata index was less than 50 (see Table III). By the techniques described (see *Methods* section) unifocalization was achievable in all patients from a midline approach.

The significant postoperative events were phrenic nerve palsy in three of our patients and severe episodic bronchospasm in three patients (two of these also had phrenic nerve palsy). These complications tended to occur in the younger patients. With proper attention to the phrenic nerve, it should be possible to completely avoid phrenic nerve palsy. Severe bronchospasm was also seen in two patients in the experience of Puga and coworkers.[7] This is probably due to the extensive dissection and disruption of lymphatics and blood vessels around the tracheobronchial tree. It is also possible

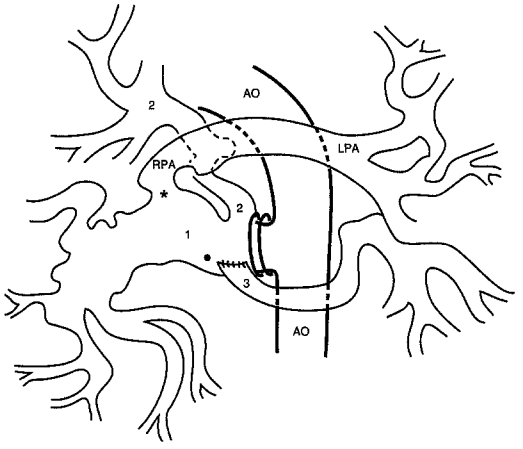

Fig. 7. Schematic representation of unifocalization in a 2.6-month-old infant who subsequently required reintervention. *1,* A large major AP collateral supplying the entire right lower and middle lobes. *Site of communication of the large collateral *1* with the true right pulmonary artery. This communication later required balloon dilation. *2,* Major AP collateral supplying the right upper lobe. *3,* Major AP collateral supplying the left lower lobe. The large collateral *(1)* was ligated flush with the aorta. Collateral *3* was disconnected from the aorta and anastomosed end to side to collateral *1*. Unifocalization was thus achieved. ●, Site of occlusion of collateral *3* at its anastomosis with collateral *1*.

that the autonomic nerve balance is affected because of the dissection. A bronchoscopic examination in one of our patients showed diffuse severe inflammation of tracheobronchial mucosa, lending validity to the concept of thermal, ischemic, or mechanical injury to the tracheobronchial tree. None of the patients had prolonged pleural or pericardial effusions. There were no neurologic events.

Younger age, longer times of cardiopulmonary bypass, and greater number of collaterals have been reported to be associated with increased mortality.[6, 11] In our experience 40% of patients were younger than 6 months of age and 50% were 9 months of age or younger. Despite the small number of patients there was no indication that young age, the number of collaterals, or the bypass time added to the postoperative morbidity or mortality.

Even in the occasional patient in whom the VSD has to be left open because of small size of the collaterals, it is advantageous to unifocalize in one stage and establish antegrade blood flow from the right ventricle into the reconstructed pulmonary arterial system. This will establish a more physiologic blood flow pattern, may encourage collateral growth, and may prevent loss of collateral-supplied segments of lung. Another significant advantage of this approach would be in providing an easy access to the interventional cardiologist for balloon angioplasty of the unifocalized collaterals and pulmonary arteries as suggested by others.[21] Maintenance of VSD patency may be warranted in some patients after unifocalization when patient referral is delayed. In our only patient without VSD closure, referral occurred at 5 years, with a history of several years of profound cyanosis (average arterial oxygen saturation in the 60% range). Even though we did not close the VSD the calculated pulmonary/systemic flow ratio at the follow-up catheterization after complete unifocalization was 1.4 to 1.8:1.0, indicating the closure might have been possible even in this setting. In this patient repair at an earlier age might well have allowed a much greater physiologic margin for simultaneous VSD closure, at a time when development of collateral stenosis had not led to subsequent distal collateral hypoplasia. This is supported by the following findings. Review of this patient's angiogram performed at 10 months of age revealed that the indexed total collateral cross-sectional area was 135 mm^2/m^2 whereas the same value was 74 mm^2/m^2 on the later angiograms at 4.2 years of age, before unifocalization.

One late death occurred in this series (at 13 months after repair). This patient also had a number of complications. At 2.6 months of age she underwent complete repair. This was followed by development of a right ventricular outflow tract pseudoaneurysm, which was repaired with conduit replacement at 7 months age. The cause of this pseudoaneurysm was not apparent. The pRV/pLV was acceptable at 0.65. It is likely that technical factors at the proximal anastomosis or homograft degeneration played a role. After this, occlusion of the unifocalized left lower lobe collateral artery was noted at follow-up—the only occlusion in 40 unifocalized vessels in this series. This complication was probably caused by a tactical error in this reconstruction. This patient was the first in this series and our approach was evolving. The situation is depicted in Fig. 7, which shows the postrepair collateral structure. Of note, the left lower lobe collateral was unifocalized to the right lower lobe collateral with the vessel crossing the midline to the left lower lobe. We believe, in retrospect, that unifocalization of this collateral by anastomosing it to the true left pulmonary artery would have provided a more durable reconstruction. This patient subsequently did have

this revised reconstruction at 13 months of age through a left thoracotomy. Although this procedure was performed uneventfully, the patient died of multiorgan failure resulting from a total body ischemic injury incurred when she had a cardiopulmonary arrest caused by delayed recognition of a compromising pneumothorax.

Ideal age of repair of this lesion is unknown. Our present approach involves the following: If the patient has well-balanced physiology, we prefer to perform this procedure between 3 and 6 months of age. However, if the patient is severely cyanotic or overshunted, repair is feasible at an even earlier age (see Table I). The advantages of early one-stage repair are numerous. Early normalization of cardiovascular physiology and correction of cyanosis are achieved. Protection against pulmonary hypertension related to high flow either through collaterals or through systemic shunts is accomplished. The number of operations is reduced. The use of nonviable material in the periphery of the lung is completely eliminated in the great majority of cases. The number of patients who can undergo complete repair is likely to be enhanced. Finally, the natural history of collateral stenosis may well be favorably affected by normalizing hemodynamics.

There are, however, some disadvantages to earlier repair. Pulmonary morbidity in the form of phrenic nerve injury and bronchospasm appear to be more likely in younger patients.

To conclude, complete unifocalization of all sources of pulmonary vascular supply and intracardiac repair of pulmonary atresia with VSD and major AP collaterals can be accomplished by a midline one-stage approach with acceptable mortality. There is little postoperative cardiac morbidity. However, proper attention to the phrenic nerve and meticulous dissection around the tracheobronchial tree are necessary to avoid the significant occurrence of pulmonary morbidity, especially in young patients. Midterm follow-up reveals excellent hemodynamics and functional status. Balloon angioplasty when required was accomplished with good results. In several instances in which late catheterization has been performed, growth of narrowed collaterals has occurred precluding the need for balloon angiolasty. In one case a lung scan has corroborated this observation. An earlier approach allows the greatest number of lung segments with healthy pulmonary microvasculature to be recruited without the need for nonviable conduits.

We speculate that early unifocalization and intracardiac repair will normalize hemodynamics and eliminate shear forces within the collaterals themselves. This may also be beneficial in preventing progressive stenosis and occlusion in the collateral vessels; that is, these collaterals may be "innocent bystanders" that react to the abnormal hemodynamics of a systemic–pulmonary artery connection by developing myointimal hyperplasia. It is also possible that future operations would be limited, in a number of cases, to central conduit changes only. As a cautionary note, we should emphasize that the long-term fate of collaterals remains uncertain. This will be a critical factor determining the late outcome of all unifocalization procedures. The validity of the approach to these complex defects outlined here is supported by the available follow-up. However, conclusive support will be achieved only with greater numbers of patients and long-term follow-up. The overall suboptimal outcome with other approaches in many patients, however, warrants continued exploration of the benefits of early one-stage unfocalization and complete repair.

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Discussion

Dr. Julie A. Swain *(Las Vegas, Nev.).* Speaking as a heart surgeon who treats adults, I think that technically this is an impressive display of results and technical ability in these very challenging cases.

Dr. Hanley. It is partly a matter of having a high tolerance for tedium as well as strictly following a plan of dissecting and controlling every single one of these collaterals before cardiopulmonary bypass. As mentioned, some of the reconstruction can be accomplished before bypass, but much of it does require bypass. This obviously takes time; however, if a warm calcium-supplemented blood prime is used for cardiopulmonary bypass, the heart remains vigorous and cardiac morbidity is minimal. Neurologic morbidity was nonexistent in this series.

Dr. G. Stellin *(Padova, Italy).* I have two questions: First, a staged repair has been undertaken in this lesion in the past because of difficulties in identifying the collateral vessel through a midline sternotomy. A right or left thoracotomy has been used to reimplant collateral vessels onto the native pulmonary artery system. Did you have any problems in isolating all of the collateral vessels through a midline sternotomy? Second, have you transplanted all of the collaterals, even those with a certain dual supply to the lungs?

Dr. Hanley. Any collateral above the diaphragm can be accessed through a median sternotomy by opening both pleura completely; the midline sternotomy incision has to be somewhat extended and the sternum widely separated. Entering both pleural spaces, and sequentially bringing each of the lungs out, allows one to gain exposure from the diaphragm to the aortic arch. The other important technique that I want to emphasize involves dissection posteriorly in the midline below the trachea but above the roof of the left atrium directly through the transverse sinus. This allows access to collaterals that are high up under the hilar regions and also provides an avenue to bring these collaterals in the most direct fashion into the unifocalization process without having to add nonviable conduits.

We were able to access all 40 collaterals in this series through this approach.

If the patient has had a previous operation, as two of our patients did, the situation is much more difficult. Therefore, I would like to emphasize that this operation should be done as the first operation. Additionally, the exposure is easier in smaller infants whose pleural spaces are less deep. Consequently the collaterals are not as far away from the surgeon.

Your other question was whether one should unifocalize all of the vessels. Again, I think the principle that every collateral should be unifocalized is very important. Some individuals follow the principle that if a segment of lung receives blood supply from both the true pulmonary artery and a collateral, ligation of the collateral should be performed. The approach that I have taken in this situation is to bring as much native tissue together by doing long onlays and long side-to-side anastomoses to build up

as much natural tissue as possible. If a large collateral supplies lung that is also supplied by the true pulmonary artery, I will use that collateral in every case to maximize using natural tissue in the conduit portion of the pulmonary arteries.

Dr. Stellin. Do you use deep hypothermia for all of your work? I am not referring to circulatory arrest.

Dr. Hanley. About half of the vessels are unifocalized before the start of bypass. That includes not only identifying them but then picking them off, one by one, and reconstructing them. The pulse oximeter value drops as each collateral is taken off, and it is possible to proceed until the patient is just short of being compromised by low oxygen saturation. Then we use a warm, partial bypass just to supply oxygen, allowing the heart to continue to beat, and then complete the unifocalization. The patient is then cooled to 25° C and the crossclamp is placed. The intracardiac repair is performed and a few of the more difficult unifocalizations may also be performed.

Dr. John L. Myers *(Hershey, Pa.).* We have had some experience with unifocalization through a midline sternotomy, although we have not progressed to complete repair. However, I certainly believe that the approach through a midline sternotomy is reasonable, particularly in the smaller infants. That approach to the descending aorta behind the aorta and behind where the pulmonary artery confluence should be allows you to see and get control of most of these collaterals. We have unifocalized them and either shunted the area that has been unifocalized or have created a right ventricular–pulmonary artery confluence conduit without closing the VSD. Your results give us additional support for proceeding with complete repair and closing the VSD.

Dr. Davis C. Drinkwater, Jr. *(Los Angeles, Calif.).* I have three questions. Do I understand correctly that you snare the collaterals before bypass to maintain an adequate perfusion pressure? Have you already obtained control of the collaterals before starting bypass? Is this always possible without destabilizing the patient?

Dr. Hanley. Yes, that's correct. At the time that we start bypass I think it is critical to have control of essentially every collateral. This is critical to adequate perfusion during bypass, especially brain perfusion.

Dr. Drinkwater. My second question is related to a possible advantage of unifocalization at a later age through the left or right thoracotomy in achieving a very large anastomosis. We believe that we can get two to three times the size of the diameter of the collateral, with potential for growth. Do you use any techniques in terms of suturing techniques and material for the future potential growth of the anastomosis in your one-stage procedure in the very young infant?

Dr. Hanley. The greatest potential for growth is achieved by getting tissue-to-tissue anastomoses wherever one can. If a previous unifocalization has been performed through a thoracotomy, there is essentially a frozen hilum, and when one approaches the hilum through the midline to do the complete repair, it is impossible to bring the vessels up to central mediastinum. It is then necessary to go down to the hilum with artificial conduits. This is a very acceptable approach in an 18- or a 20-year-old patient, one of those highly selected patients who have just the right amount of collateral stenosis such that they have had enough flow into their lungs that the alveoli and small vessels have grown adequately, but they are not in trouble from pulmonary vascular obstructive disease. Artificial conduits are acceptable in these patients because growth potential is not an issue. When there is no need for growth, it is not so critical to get these tissue-to-tissue anastomoses. However, if one were to observe a cohort of patients with this lesion from birth, an extremely small percentage will reach adulthood and still have repairable lesions. Therefore a wait and see approach is not likely to achieve the most good. Similarly, a delayed staging approach will also have substantial dropout of candidates, with probably less than 30% of all patients receiving a good repair. If these patients are treated in infancy, all of the tissues can be brought together such that there is no artificial material other than the central conduit. This should maximize the number of complete repairs and provide growth potential for all segments of the pulmonary arterial tree except the central conduit. With this approach we have shown that there is a 90% likelihood of complete repair. The long-term outcome remains unknown but will depend substantially on the growth potential of the collateral vessels, as is the case with any approach to this lesion.

Dr. Drinkwater. My third point is that we have good exposure through a "clamshell" incision for the bilateral lung transplant. Could this be used to advantage in this procedure to improve access to the posterior mediastinum?

Dr. Hanley. That is an excellent suggestion. We have not used this incision, but I would imagine that it would be equally efficacious, if not more efficacious, in allowing us to achieve the necessary exposure.