SURGERY FOR CONGENITAL HEART DISEASE

UNIFOCALIZATION FOR PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND MAJOR AORTOPULMONARY COLLATERAL ARTERIES

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To extend the indications for corrective operation in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries, surgical procedures were done to unify the blood sources for pulmonary perfusion. Since December 1985, 50 patients have undergone unifocalization at ages from 2 months to 26 years with a mean of 6 ± 7 years. In total, 84 staged unifocalization procedures and 5 other palliative procedures were done in 49 patients. These included several operative procedures: simple ligation of major aortopulmonary collateral arteries in 8; pulmonary angioplasty in 29 including reconstruction of the pulmonary arterial tree by direct anastomosis or interposition between the central pulmonary arteries and the intrapulmonary arteries; construction of artificial central pulmonary arteries with use of a xenograft pericardial tube graft in 36 with no native central pulmonary arteries detected; and construction of supplemental central pulmonary arteries also with use of a pericardial tube graft in 10. The pericardial tube graft, if used, was anastomosed to the intrapulmonary arteries on one end and connected to a prosthetic tube on the other end so as to perfuse the reconstructed pulmonary arteries. The anastomosis was made inside the lung through the divided interlobar fissure. Five patients died after operation among those undergoing these 89 preparative operative procedures. Deaths were related either to bleeding caused by anticoagulation therapy administered to prevent thrombosis within the xenograft pericardial tube graft used or to progressive congestive heart failure as a result of an excessive amount of pulmonary blood flow. Twenty-six patients have undergone intracardiac repair after previous unifocalization. In 16 patients the artificial central pulmonary arteries surgically constructed were connected to each other and then an external conduit was placed. In another patient, intracardiac repair and unifocalization could be concomitantly achieved via a median sternotomy. The right ventricle to left ventricle systolic pressure ratio immediately after intracardiac repair in 27 patients ranged from 0.24 to 0.91 with a mean of 0.54 ± 0.17 . One patient (4%) died shortly after intracardiac repair because of thrombosis within the pulmonary arteries. Postoperative catheterization showed that pulmonary vascular resistance was correlated significantly with the number of pulmonary vascular segments functioning rather than with the condition of the central pulmonary arteries. We conclude that surgical unifocalization is a feasible procedure before subsequent intracardiac repair, even in patients with critically hypoplastic or absent central pulmonary arteries. (*J Thorac Cardiovasc Surg* 1996;112:392-402)

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The precise diagnosis and successful surgical interventions in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary (AP) collateral arteries have been documented.¹⁻⁸ The operative procedures for unifocalizing blood supply to the lung, however, markedly vary from one another, and determination of the optimal surgical approach to this complicated malformation with abnormal pulmonary arterial tree, particularly in patients with vestigial sizes of the central pulmonary arteries, remains a matter of controversy. Furthermore, the prognoses in the long term after these surgical interventions have yet to be unequivocally determined.

Since December 1985, we have striven to establish the surgical unifocalization of pulmonary blood supply in a series of patients. In the present study, we attempted to analyze the surgical results and prognoses in these patients from the perspective of the pulmonary arterial morphologic features and hemodynamic data derived from clinical examinations.

Methods

Patients. Between December 1985 and March 1993, 50 of 52 consecutive patients with pulmonary atresia, ventricular septal defect, and major AP collateral arteries underwent surgical unifocalization as initial operative procedures aiming toward intracardiac repair at the National Cardiovascular Center in Osaka. One of the two patients excluded from this series was a 24-year-old woman with severe aortic insufficiency who initially underwent aortic valvular replacement with no further procedures. The other was a small infant with severe hypoxia, in whom the central pulmonary arteries and all the intrapulmonary arteries were vestigial. In this particular patient only construction of a systemic-pulmonary shunt could be done.

Of the 50 patients in our present series, 31 were male and 19 female. Ages at initial unifocalization ranged from 2 months to 26 years old, with a mean of 5.9 ± 6.7 years (Fig. 1). Nineteen patients (38%) had a right aortic arch. In 10 patients, some palliative procedures such as construction of a systemic-pulmonary shunt had been done before 1985 when unifocalization began to be done at our institute. These previous procedures were not taken into account in the analyses.

Morphologic features of pulmonary arteries. The structural features of the pulmonary arterial vasculature and major AP collateral arteries were determined by panaortography and selective injection of contrast media into each major AP collateral artery before operation, as well as by injection into the surgically constructed shunt after operation.

Central pulmonary arteries. The central pulmonary arteries could be preoperatively identified in 42 patients. In contrast, these arteries were likely lacking in 8 patients. Of

the 42 patients with the central pulmonary arteries, 3 patients had nonconfluent central pulmonary arteries and another patient had only a unilateral portion of the central pulmonary arteries.

In our strategies, the minimal Nakata's index for successful repair of tetralogy of Fallot has been considered to be about the value of 120.⁹ We had classified our patients who had an index value of 120 or greater into the "medium sized" group ($n = 11$). Another 18 patients had a pulmonary arterial area index less than 120, but the absolute value for the diameter of the central pulmonary arteries was still greater than 2 mm (the "small" group). The central pulmonary arteries were diminutive with a diameter of less than 2 mm at any portion in 13 patients (the "vestigial" group). The 8 patients with no detectable central pulmonary arteries were unified under the heading of the "absent" group. Eventually, each patient was classified into one of these four groups according to the angiographic findings in the central pulmonary arteries as shown in Fig. 2.

AP collateral arteries. According to previous reports,^{5,6} a major AP collateral artery was defined as a large collateral vessel originating proximally from the aorta or its major branch and distally connected with the intrapulmonary arteries at the hilum. Collateral arteries secondarily developed, such as either the native bronchial arteries or the vessels between the intercostal arteries and the parenchyma of the lung, were distinguished from major AP collateral arteries. The overall number of major AP collateral arteries detected in all the 50 patients was 160, with the number per patient ranging from 1 to 6 with a mean of 3.3 ± 1.1 . Of these 160 major AP collateral arteries, 139 (87%) originated from the descending aorta, 20 (12%) from the subclavian artery, and 1 (1%) from the abdominal aorta. Irrespective of the origins, 115 major AP collateral arteries (72%) were considered to be an independent source of the blood supply for some pulmonary segments; this was seen in 44 patients (88%). In such circumstances, communications of blood flow were angiographically lacking between the isolated pulmonary segments perfused via a major AP collateral artery and the other segments. In contrast, 45 (28%) proved to perfuse some pulmonary segments and were incorporated with other intrapulmonary arteries; that is, the segments were dually supplied.

Surgical method

Unifocalization and other palliations. The surgical procedures for unifocalization of pulmonary blood supply were done in staged operations through lateral thoracotomy incisions. In total, 84 unifocalizations were accomplished. Among these, construction of a systemic-pulmonary shunt was simultaneously done in 75 operations with use of a knitted Dacron tube graft of, in most cases, 5 mm in diameter. The major AP collateral arteries were extensively dissected from the origins on either the aorta or the subclavian artery to the hilum of the lung. Intrapulmonary arteries were also exposed inside the lung through divided interlobar fissures. After the precise anatomic features of the intrapulmonary arteries, the central pulmonary arteries, and the major AP collateral arteries were determined, and taking the patterns of communications between them into account, the operative procedure to be used for

Fig. 1. Number of patients (Pt.) and age at first operation.

Fig. 2. Patients were classified into one of four groups according to morphologic features of central pulmonary arteries. The medium-sized group represents patients with adequate sized central pulmonary arteries greater than minimal size for successful intracardiac repair. The small group comprises patients with hypoplastic central pulmonary arteries that are greater than 2 mm in diameter. Vestigial group comprises patients having central pulmonary arteries less than 2 mm in diameter. In absent group, central pulmonary arteries could not be identified angiographically. Brighter gray indicates central pulmonary arteries and darker gray represents intrapulmonary arteries for which blood is supplied mainly via major AP collateral arteries.

reconstruction of the pulmonary arterial tree was eventually decided.

The variations in techniques for unifocalization in patients with central pulmonary arteries are shown in Fig. 3. Ligation of major AP collateral arteries at their origins concomitant with construction of a modified Blalock-Taussig shunt was the simple procedure of choice done eight times in eight patients; the central pulmonary arteries were considered to provide an adequate channel for pulmonary perfusion. Angioplasty with a patch was required six times because of the restrictive nature of the communications between the central pulmonary arteries and the intrapulmonary arteries. In 14 patients in whom such communications were lacking or extremely hypoplastic, the intrapulmonary arteries were directly anastomosed; this was done 17 times. If the intrapulmonary arteries connected to the major AP collateral arteries were located just behind the pulmonary veins or the bronchi, the direct anastomosis was accomplished by

means of dividing such major AP collateral arteries and mobilizing them from the dorsal side of the hilum to the interlobar space. An extrapulmonary portion of the major AP collateral arteries was used as necessary for direct anastomosis. If direct anastomosis did not prove feasible, either a segment of the azygos vein or a heterologous pericardial tube graft was interposed for reconstruction of the pulmonary arteries; this modification was done six times in six patients.

In patients with small central pulmonary arteries, a supplemental central pulmonary artery made of a heterologous pericardial tube graft was placed. In this procedure, intrapulmonary arteries were divided from the major AP collateral arteries and anastomosed to the xenograft pericardial tube graft. Two minor modifications were used to deal with the proximal end of the tube graft, as shown in Fig. 4. Side-to-side anastomosis was done seven times, and construction of "double central pulmonary arteries" three times.

Fig. 3. Basic procedures to unifocalize intrapulmonary arteries in patients with central pulmonary arteries. **a**, Ligation of major AP collateral artery or coil embolization; **b**, repair of stenotic communication of intrapulmonary arteries with or without patch; **c**, direct anastomosis between intrapulmonary arteries; **d**, interposition between intrapulmonary arteries with azygos vein graft or pericardial tube graft (*roll*).

Fig. 4. Construction of supplemental central pulmonary artery with xenograft pericardial tube in patients with slender central pulmonary arteries. In *upper left*, proximal side of tube is anastomosed to central pulmonary artery in side-to-side fashion. *Upper right* shows “double central pulmonary arteries” (CPA). *Lower illustrations* show how to reconstruct central pulmonary arteries at time of intracardiac repair. **a**, Tube was connected to external conduit after enlargement of central pulmonary arteries with use of patch. **b**, Tube was unified together with native central arteries.

Construction of entirely new central pulmonary arteries with use of a heterologous pericardial tube graft was done 36 times in 24 patients with vestigial or absent central pulmonary arteries. Dissected intrapulmonary arteries were anastomosed to one end of the tube graft to unify the independent pulmonary vascular segments. The other end of the tube graft was closed and fixed to the mediastinum,

and a prosthetic tube graft was interposed there between either the subclavian artery or the ascending aorta and the tube graft to supply blood flow to the lung (Figs. 5 and 6).

In patients with a xenograft pericardial tube graft, anticoagulation was begun by infusion of heparin sodium from the first postoperative day to maintain the activated clotting time at about 150 seconds. Heparin infusion was

Fig. 5. Construction of artificial central pulmonary artery with xenograft pericardial tube (*roll*) was done in patients with critically hypoplastic or absent central pulmonary arteries. All intrapulmonary arteries were anastomosed to tube, and proximal end of tube was closed and fixed to mediastinum. SPS, Systemic-pulmonary shunt.

Fig. 6. Pulmonary arterial angiography before (*left*) and after (*right*) unifocalization in patient with absent central pulmonary artery. *Pericardial roll*, Pericardial tube graft.

discontinued when oral administration of warfarin potassium and dipyridamole was feasible. Obstructions within the tube graft caused by thrombosis have occurred three times, although thrombi could be successfully removed.

In addition to these procedures, central palliation that could facilitate growth in small or vestigial central pulmonary arteries was done in five patients, as an initial palliative procedure in three and the third palliative procedure in two. Each procedure was done through a median sternotomy. In four children, the right ventricular outflow tract was created with use of a heterologous pericardial tube graft. In a 2-month-old infant, the minute central pulmonary arteries, which were approximately 2 mm in diameter, were directly anastomosed to the ascending aorta.

Intracardiac repair. Twenty-six patients underwent definitive repair after the staged unifocalizations, whereas one patient underwent intracardiac repair concomitant with unifocalization through a median sternotomy without any previous procedures. Of the 27 patients, 17 were male and 10 female. Age at repair ranged from 1 to 24 years (mean plus or minus SD, 7 ± 5 years). The number of surgical procedures done before the intracardiac repair was 1 to 3 per patient, with a mean of 1.8. The preparative procedures included 2 central palliations and 45 unifocalizations (4 by simple ligation of major AP collateral arteries, 14 by arterioplasty of the pulmonary arterial tree

*Standard deviation.

Fig. 7. Artificial central pulmonary arteries were connected to each other with use of another pericardial tube graft (*roll*) at time of intracardiac repair. The interposed tube graft was placed posterior to aorta (**a**) in one patient and anterior to aorta (**b**) in nine. *SVC*, Superior vena cava; *Ao*, aorta.

Fig. 8. Right ventriculography (*left*) and pulmonary arteriography (*right*) after definitive repair in patients shown in Fig. 6. *Pericardial roll*, Pericardial tube graft; *RV*, right ventricle.

including 7 by direct anastomosis and 3 by interposition of the intrapulmonary arteries, 5 by creation of a supplemental central pulmonary artery with use of a tube graft, and 22 by construction of the central pulmonary artery with use of a tube graft). The interval between the most recent preparative operation and the intracardiac repair was 1 month to 5

years (mean plus or minus SD, 13 ± 14 months). In two patients, embolization of residual major AP collateral arteries with coils was done by means of a transcatheter technique just before the intracardiac repair.

Among the 10 patients with medium-sized central pulmonary arteries, the right ventricular outflow tract was

Fig. 9. In 20 patients, ventricular septal defect was closed with nonfenestrated patch (*left*). Fenestrated patch bearing valvular structure was used in six patients (*middle*). Several sutures were placed for closing fenestration from outside heart. In five of six patients, fenestration could be closed because systolic pressure of right ventricle was relatively low. Fenestrated patch bearing no valve was used in one patient (*right*). Ao, Aorta; RV, right ventricle; LV, left ventricle; IVS, interventricular septum.

reconstructed with a handmade external conduit containing a valve with three leaflets in seven patients and with use of a patch with a monocusp in three patients.

The surgically constructed central pulmonary arteries formed by tube grafts were connected to each other interposing another tube graft to reconstruct the confluence between the right and the left pulmonary arteries. Reconstruction of the right ventricular outflow tract was accomplished with an external conduit containing a valve (Figs. 7 and 8). In the majority of the patients, the maneuver for reconstruction of the confluent pulmonary arteries could be done before cardiopulmonary bypass was started. The interposed tube graft was placed anterior to the aorta in nine patients and posterior to the aorta in one.

The method of intracardiac repair is shown in Fig. 9. In 20 patients, the ventricular septal defect was entirely closed with use of a nonfenestrated patch. A fenestrated patch bearing a valvular structure was used in six patients, and a fenestrated patch with no valvular device was used in another patient. In these patients, it was estimated before operation that systolic pressure of the right ventricle would remain excessively high after intracardiac repair because of the high pulmonary resistance. Intraoperatively, however, the fenestration could be closed by fixing the valve to the patch after cardiopulmonary bypass was discontinued in five patients. Eventually, the fenestration was left open in two patients, one with a valve and the other without.

Results

Preparative operations. A flow chart of overall courses and results in all patients treated with the aim of eventual intracardiac repair is shown in Fig.

Fig. 10. Results in all patients in present series shown schematically. PA, Pulmonary atresia; VSD, ventricular septal defect; MAPCA, major AP collateral arteries; pts, patients; UF, unifocalization; CP, central palliation.

10. In the course of the overall 89 preparative procedures done, five patients (10%) died before discharge from the hospital. Three deaths were related to the subacute onset of hemothorax, with two of these accompanied by cardiac tamponade. Another patient died of massive esophageal bleeding that occurred suddenly during mechanical ventilation in the intensive care unit. A postmortem study demonstrated ulceration and perforation at the posterior wall of the esophagus, with the lesion being located where the nasogastric tube by adjacent

to the abnormal course of the right subclavian artery, which had an aberrant origin and which was dilated markedly because of a previously constructed systemic-pulmonary shunt. The remaining one patient who underwent central palliation subsequent to bilateral unifocalizations, despite having done well immediately after the procedure, died suddenly probably because of a circulatory pulmonary-systemic mismatch induced by an excessive amount of blood flow to the lung.

Five patients have been dropped from the present series: in three patients the condition proved unsuitable for repair because of either the small number of pulmonary vascular segments (in 2) or the structural features of the ventricles (in 1), and the other two patients died in the intermediate term of respiratory infection.

Intracardiac repair. Intracardiac repair was accomplished with cardiopulmonary bypass lasting from 71 to 467 minutes (mean plus or minus SD, 186 ± 81 minutes), including an aortic crossclamp time of 32 to 155 minutes (mean, 58 minutes). Postoperative pressure studies in the operating room demonstrated a right ventricle/left ventricle systolic pressure ratio ranging from 0.24 to 0.91 (mean plus or minus SD, 0.54 ± 0.17), a mean pulmonary arterial pressure of 10 to 35 mm Hg (21 ± 7 mm Hg), and a pressure gradient between the right ventricle and the pulmonary arteries of 0 to 35 mm Hg (10 ± 9 mm Hg).

One patient (4%) died suddenly after intracardiac repair as a result of pulmonary emboli on postoperative day 8. Eleven pulmonary vascular segments had been counted before the operation in this particular patient, and the postoperative right ventricle/left ventricle systolic pressure ratio was measured at a value as high as 0.91; these findings were the worst among all the patients. Five patients (19%) have died in the intermediate term. One patient died of gastrointestinal bleeding 1 month after repair. Another patient had repeated bronchial bleeding and died 3 months after repair. The third died of intractable mediastinitis caused by methicillin-resistant *Staphylococcus aureus* infection 4 months after repair. The fourth had been doing well, but this patient's condition deteriorated 5 months after repair. Viral myocarditis was suspected to be the cause of fatal ventricular dysfunction in this patient. The last patient had occasional difficulty in breathing because of bronchial stenosis caused by compression by the constructed central pulmonary artery behind the aorta. Respiratory infection was

Fig. 11. Relationship between number of pulmonary vascular segments (Seg.) counted on preoperative angiography and pulmonary vascular resistance (Rp) calculated at time of postoperative catheterization. There was significant adverse correlation ($p < 0.01$) between them. CPA, Central pulmonary artery.

the cause of death in this patient 3 years after intracardiac repair. In these five patients, the postoperative right ventricle/left ventricle systolic pressure ratio was 0.24 to 0.79 (mean, 0.58), and the hemodynamic condition was initially stable.

Postoperative catheterization was done in 20 patients. The interval between intracardiac repair and the catheterization ranged from 1 to 64 months with a mean of 17 ± 15 months. Four patients had a right ventricle/left ventricle systolic pressure ratio higher than 0.8. Of these four patients, one with moderately progressed stenosis within the external conduit and with mild pulmonary hypertension underwent replacement of the conduit 6 years after intracardiac repair. Another patient had markedly elevated pulmonary arterial pressure, and the other two patients had both moderate pulmonary hypertension and a moderate pressure gradient across the right ventricular outflow tract. Residual interventricular shunts were either trivial or absent in all patients but one who had a fenestrated patch, in whom the pulmonary/systemic flow ratio was calculated at 1.14. In the patient with the fenestrated patch bearing a valvular structure that was left unfixed, ventriculography 1 month after operation showed spontaneous closure of the fenestration. As is shown in Fig. 11, pulmonary vascular resistance calculated at the time of postoperative catheterization was adversely correlated with the number of pulmonary vascular segments that were counted on preoperative angiography ($p < 0.01$). The size of the central pulmonary

arteries showed no obvious correlation to postoperative values of pulmonary resistance.

The most recent clinical features in 21 patients surviving intracardiac repair show that the patients are doing well with good physical development and freedom from episodes of arrhythmia. Exercise tolerance has also improved in all patients but one with severe pulmonary hypertension.

Discussion

Many surgeons have previously reported improved results of intracardiac repair for pulmonary atresia with ventricular septal defect without major AP collateral arteries,^{10,11} even in the setting of hypoplastic central pulmonary arteries.^{12,13} The quantitative criterion for successful repair in their patients was determined by the size of the central pulmonary arteries.⁹ Quantitative criteria, as well as qualitative ones, for successful repair in patients with major AP collateral arteries, however, have yet to be established, particularly in patients with independent sources for blood supply to some segments of the lung.

The structural features of major AP collateral arteries have been extensively studied by some cardiologists,³⁻⁶ and the feasibility of surgical intracardiac repair of pulmonary atresia, ventricular septal defect, and major AP collateral arteries has been suggested in the past decade if the abnormal sources of pulmonary perfusion via major AP collateral arteries could be unified and used as parts of the pulmonary arterial tree.¹⁻³ In recent years, several surgeons have attempted to achieve "unifocalization" as preparative surgical steps aiming toward subsequent intracardiac repair of cardiac malformations coexisting with major AP collateral arteries. Although this procedure can provide disappointing results,¹⁴ other surgeons have demonstrated a vast improvement in their results in a series of patients.¹⁵⁻¹⁸ In the initial era, unifocalization was considered feasible in a limited group of patients with central pulmonary arteries that were not extremely small by connecting the intrapulmonary arteries supplied by major AP collateral arteries to the central pulmonary arteries.¹⁴ In patients with vestigially small central pulmonary arteries or in those with no formation of the central pulmonary arteries, the operative methods for creating the artificial central pulmonary arteries with use of a xenograft pericardial tube graft¹⁵ or a prosthetic tube^{16,19,20} have provided the alternative surgical approach in preparation for future definitive repair.

How to manage the small sizes of the pulmonary

arteries remains a matter of controversy. Some surgeons attempted to promote growth of the pulmonary arteries by constructing the channel from either the right ventricular outflow tract or the ascending aorta to the hypoplastic central pulmonary arteries as an initial palliative procedure.^{12,13,17} Their principles are attractive, and could be beneficial, particularly in infants with the confluent right and left pulmonary arteries perfusing greater parts of the pulmonary segments. We have basically striven to accomplish another approach in which the central pulmonary arteries are constructed entirely or supplementally with a xenograft pericardial tube graft. This procedure is seemingly advantageous in children and adolescents with hypoplastic central pulmonary arteries and major AP collateral arteries independently perfusing parts of the lungs.

On the basis of findings of our present study, the patterns of the intrapulmonary arteries, the central pulmonary arteries, and major AP collateral arteries are reasonably classified so as to determine the optimal method and timing of the initial preparative operation. The combination of high pulmonary blood flow and the presence of a certain degree of stenosis within major AP collateral arteries can be favorable and is likely the commonest variation. In such circumstances, intrapulmonary arteries are usually well developed, pulmonary vascular changes are mild or absent, and, in addition, the clinical condition of these patients is generally stable. Staged unifocalizations via lateral thoracotomies are feasible in infancy or childhood, or even in adolescence. In patients with high pulmonary blood flow and absence of stenosis within major AP collateral arteries, excessive pulmonary flow may produce unfavorable changes in the pulmonary vessels and raise the pulmonary resistance. In this circumstance, surgical treatment for reducing and controlling the amount of pulmonary blood flow in early infancy should be planned. The limitation, however, is extensive use of prosthetic materials, which would not be justified because of lack of growth potential. The reversed combination of low pulmonary blood flow and the presence of stenosis within major AP collateral arteries is also difficult to treat, because the clinical condition is generally unstable because of hypoxia. An initial procedure should be planned for augmenting the amount of pulmonary blood flow. In such patients, particularly in small babies with tiny central pulmonary arteries, the construction of a channel from the ascending aorta to the pulmonary

artery might be the best option.¹⁷ The most deleterious condition is undoubtedly low pulmonary blood flow even though no stenosis is found within major AP collateral arteries. In this situation the intrapulmonary arteries must be severely hypoplastic, although obstructive pulmonary vascular disease may be progressive. Furthermore, the clinical status is usually unstable, and attempts to increase the amount of pulmonary blood flow are frequently unsuccessful.

In our series of patients, unifocalization in older children or adolescents appeared rather successful. This is probably because they possessed the combination of high pulmonary blood flow and stenotic lesions within major AP collateral arteries. Furthermore, patients with adverse risk factors had already been excluded, having died earlier because of critical hypoxia or intractable congestive heart failure. However, recent reports suggest that good surgical results can be achieved in such patients properly selected.¹⁶

Obviously, the characteristic problems in patients with major AP collateral arteries lie not only in either regionally unbalanced blood perfusion to the lung or abnormal growth of intrapulmonary arteries, but also in the presence of various pulmonary vascular resistance within the lungs.⁴ If obstructive pulmonary vascular changes would regionally progress, unifocalization of the origins for blood flow to the lung should produce unbalanced perfusion within the lung after operation. Indeed, some pulmonary vascular segments disappeared angiographically in some of our patients after unifocalizations. To prevent unbalanced pulmonary perfusion and to salvage the pulmonary vascular segments as much as possible, earlier procedures for normalizing the pulmonary circulation are mandatory.¹⁸ Of course, from the technical aspect, it is essential to reconstruct the intrapulmonary arteries with no stenosis at the sites of anastomoses. To avoid surgical obstruction, we emphasize that intrapulmonary arteries should be extensively exposed inside the lung through the divided interlobar fissure.

The finding that the number of functioning pulmonary vascular segments was adversely correlated to pulmonary resistance after definitive repair was comparable to similar results previously reported in patients with pulmonary atresia and ventricular septal defect in whom unifocalization was not established.²¹ On the basis of this finding, our policy to salvage pulmonary segments as much as possible by means of earlier unifocalizations is well justified.

The optimal method and the optimal timing for unifocalizations, as parts of the staged repair of pulmonary atresia with ventricular septal defect and major AP collateral arteries, should be considered in each individual patient according to the morphologic and hemodynamic features of the pulmonary circulation.

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