ONE-STAGE MIDLINE UNIFOCALIZATION AND COMPLETE REPAIR IN INFANCY VERSUS MULTIPLE-STAGE UNIFOCALIZATION FOLLOWED BY REPAIR FOR COMPLEX HEART DISEASE WITH MAJOR AORTOPULMONARY COLLATERALS

Christo I. Tchervenkov, MD^a
Gary Salasidis, MD^a
Renzo Cecere, MD^a
Marie J. Béland, MD^b
Luc Jutras, MD^b
Marc Paquet, MD^b
Anthony R. C. Dobell, MD^b

Background: Patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries have traditionally required multiple unifocalization staging operations before undergoing complete repair. Recently, the feasibility of a single-stage unifocalization and repair was demonstrated by Hanley. In this report, we describe our experience with each approach. **Methods and Results:** Since 1989, 11 of 12 patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries have undergone complete surgical correction. The first seven patients were subjected to staged bilateral unifocalizations, with repair being achieved in six (group I). The last five patients have undergone a single-stage midline unifocalization and repair via a sternotomy (group II). Four of these were infants (2 weeks to 9 months) and one was 13 years old. All patients in group I had tetralogy of Fallot, whereas in group II three patients had tetralogy of Fallot, one patient had double-outlet right ventricle, and one patient had complete atrioventricular canal and transposition. In group I, the median age at the first operation was 43 weeks. Complete repair was performed at a median age of 3.5 years, with a mean number of 3.3 operations required. In group II, only one operation was required to achieve complete repair at a median age of 28 weeks. The postoperative right ventricular/left ventricular pressure ratio was 0.49 in group I and 0.45 in group II. One intraoperative death and one late death occurred in group I and no early or late deaths in group II. Currently, four patients in group I and all five patients in group II are alive and well. **Conclusions:** Early intervention with both surgical approaches can lead to complete biventricular repair in most patients. Because the single-stage midline unifocalization and repair can achieve a completely repaired heart in infancy with one operation, it is currently our approach of choice. (J Thorac Cardiovasc Surg 1997;114:727-37)

Patients with pulmonary atresia, ventricular septal defect (VSD), and major aortopulmonary collateral arteries (MAPCAs) have always been a formi-

dable surgical challenge. Traditionally, these patients were simply subjected to palliation, and complete repair was considered an unrealistic goal. Several groups, however, have demonstrated the feasibility of achieving complete repair with a protocol of multiple preparatory operations to unifocalize and centralize pulmonary blood flow.¹⁻⁶ A significant number of these patients, however, never reach the repair stage, because of either interim death or an inadequate distal pulmonary arterial bed. Recently, Reddy, Liddicoat, and Hanley⁷ have demonstrated the feasibility of a single-stage unifocalization and repair via a median sternotomy in a series of patients. We report on our experience at the Montreal Children's Hospital with both these approaches.

From the Division of Cardiovascular Surgery^a and the Division of Cardiology,^b The Montreal Children's Hospital, McGill University, Montreal, Quebec, Canada.

Read at the Seventy-seventh Annual Meeting of The American Association for Thoracic Surgery, Washington, D.C., May 4-7, 1997.

Received for publication May 7, 1997; revisions requested June 12, 1997; revisions received July 7, 1997; accepted for publication July 9, 1997.

Address for reprints: Christo I. Tchervenkov, MD, Director of Cardiovascular Surgery, The Montreal Children's Hospital, 2300 Tupper St., Room C-827, Montreal, Quebec, Canada H3H 1P3.

Copyright © 1997 by Mosby-Year Book, Inc.

0022-5223/97 \$5.00 + 0 12/6/84567

Table 1. Patient demographics (n = 12)

	Multiple stage (n = 7)		Single stage (n = 5)	
	Median	Range	Median	Range
Age (wk)				
First operation	43	1-228	28	2-711
Final repair	182	145-495	28	2-711
Weight (kg)				
First operation	6.4	3.4-13.1	6.3	3.4-37.4
Final operation	13.0	9.5-19.7	6.3	3.4-37.4
Sex				
Male/female	4:3		2:3	

Methods

Patients. From May 1989 to April 1997, 12 consecutive patients with pulmonary atresia, VSD, and MAPCAs have been entered in a pulmonary unifocalization and repair protocol. The patients included six boys and six girls. The demographic characteristics are summarized in Table 1. The first seven patients were subjected to a multiple-stage approach of preliminary unifocalizations, with one patient dying after a left pulmonary unifocalization. This patient will be excluded from subsequent comparisons between groups. Therefore six patients reached the reparative stage (group I). Since May 1995, the last five patients in the series have undergone a single-stage bilateral unifocalization and repair through a median sternotomy incision (group II). Of the 11 patients in whom complete repair was achieved, eight were infants at the first operation (four of six in group I and four of five in group II). Two patients were neonates, one in each group.

The diagnosis was made or suspected on the basis of echocardiography and confirmed by cardiac catheterization. The MAPCAs were identified at aortography and their anatomy was further delineated by selective angiography when possible. In the older patients, pressure measurements were made to rule out pulmonary vascular obstructive disease. The native pulmonary arteries were identified before the operation whenever possible and their size was assessed by calculating the Nakata index.⁸ An effort was made to delineate as accurately as possible the blood supply to each area of the lung and the areas of the lung with dual blood supply. Of particular importance to our surgical approach was to determine whether a MAPCA was communicating or not communicating with the native pulmonary circulation. The native pulmonary arteries were completely absent in three patients (one of six patients in group I and in two of five patients in group II). The morphologic characteristics of the MAPCAs and the native pulmonary arteries in the patients undergoing complete repair are summarized in Table II. The anatomic details of the pulmonary blood supply are schematically depicted in Fig. 1 for the six patients subjected to multistage unifocalization followed by repair (group I). Similarly, the pulmonary blood supply in the five patients undergoing single-stage midline unifocalization and repair (group II) is shown in Fig. 2. All patients in group I had tetralogy of Fallot, whereas in group II three patients had tetralogy of Fallot, one patient had double-outlet right

ventricle, and one patient had complete atrioventricular canal and transposition of the great arteries. One patient in group I had previously undergone a central aortopulmonary shunt to a diminutive main pulmonary artery via a sternotomy at another institution. This shunt had thrombosed and the patient was referred to the Montreal Children's Hospital for consideration for unifocalization and repair. The other patients had no previous operations before referral to us.

Surgical technique

Multistage unifocalization followed by repair. At the initial operation, one of the goals was to establish flow in the native pulmonary arteries to promote their growth when possible. In the first patient in the series, at 9 days of age, the initial operation was to create a confluence between the discontinuous diminutive native right and left pulmonary arteries and place a central shunt into the reconstruction. In the other six patients, the initial operation by us was a left pulmonary unifocalization via a left posterolateral thoracotomy. The left-sided approach allowed us to open the pericardium, inspect the intrapericardial native pulmonary arteries, and assess their size, confluence, and the presence or absence of a main pulmonary artery. In one patient, the main pulmonary artery was transected and sutured to the side of the ascending aorta. In the other patients, either a modified Blalock-Taussig shunt or a modified Waterston shunt (ascending aorta to branch pulmonary artery with polytetrafluoroethylene*) was performed either to the native pulmonary arteries when present or to the unifocalized reconstruction. The MAPCAs were handled in several ways:

1. End-to-side or side-to-side anastomosis to the native pulmonary artery
2. Side-to-side anastomosis to other MAPCAs
3. Beveled end-to-side anastomosis to a polytetrafluoroethylene tube
4. Ligation of adequately communicating MAPCAs to the native pulmonary arteries
5. Ligation of MAPCAs to areas of the lung receiving significant dual blood supply
6. Ligation of small MAPCAs supplying a single bronchopulmonary segment if unifocalization of that segment was too difficult

After the left pulmonary unifocalization, the same process was later repeated on the right side via a right posterolateral thoracotomy. In general, the interval between the two unifocalization procedures was several months. After the second unifocalization, the definitive repair was delayed for at least a year to allow for maximal growth of the native pulmonary arteries. In one patient, the complete repair was delayed for an additional 4 years because the parents were unwilling to accept the surgical risk until the patient's status had deteriorated significantly.

The intracardiac repair consisted of closure of the VSD in all patients, because the total number of unifocalized segments in each patient exceeded at least one lung equivalent.² All atrial septal defects were also closed,

*Gore-Tex tube, registered trade mark of W. L. Gore & Associates, Inc., Elkton, Md.

Table II. Characteristics of MAPCAs and native pulmonary arteries of patients in whom full repair was achieved

	Group I: Multiple stage (n = 6)		Group II: Single stage (n = 5)	
	Mean	Range	Mean	Range
MAPCA				
Total/patient	4.3	2.0-6.0	3.2	2.0-4.0
Ligated/patient	1.8	0.0-2.0	1.6	0.0-3.0
Transplanted/patient	2.5	0.0-4.0	1.6	1.0-2.0
LPA at presentation (mm)	3.5	0.0-5.0	1.4	0.0-4.4
RPA at presentation (mm)	3.0	0.0-4.6	3.7	0.0-7.2
Nakata index (at presentation)	69	0.0-145	70	0.0-164

LPA, Left pulmonary artery; RPA, right pulmonary artery; MAPCA, major aortopulmonary collateral artery.

whereas small patent foramina ovalia were left open. Right ventricular–pulmonary artery continuity was established with a valved pulmonary homograft of appropriate size. Native pulmonary confluence, when present, was enlarged with the distal end of the pulmonary homograft by using the opened right and left pulmonary artery branches as patches. In the absence of pulmonary artery confluence, the right and left pulmonary artery branches of the homograft were used intact to create confluence between the left and right unifocalized hilar tubes. The operations were performed with deep hypothermia (about 20° C) to be able to lower the pump flow to control the noncoronary collateral return and allow accurate hilar anastomoses. The VSD was closed with a polytetrafluoroethylene patch through a vertical right ventriculotomy. The proximal end of the homograft was sutured to the distal end of the ventriculotomy incision and augmented with a hood of bovine pericardium. Approximately half an hour after discontinuation of cardiopulmonary bypass, the right ventricular/systemic pressure ratio was recorded. This measurement was repeated 24 to 48 hours after the operation to assess the adequacy of the pulmonary vascular bed as faced by the right ventricular output. An intraoperative transesophageal echocardiogram was performed to evaluate the repair, rule out significant residual lesions, and assess ventricular contractility and valve function.

Single-stage bilateral unifocalization and complete repair. After a midline sternotomy incision, a pericardial patch was removed to be used subsequently for the outflow tract reconstruction. The MAPCAs were reached by a dissection in the posterior mediastinum, without opening the pleura. The right-sided MAPCAs originating from the descending aorta were reached by dissecting the area between the superior vena cava, the ascending aorta, and the roof of the left atrium, usually below the carina and the right main-stem bronchus. Dissection was often guided by feeling the thrill in the collaterals to confirm their location. The native right pulmonary artery was also mobilized through this exposure. After localization of the right-sided collaterals and before their takedown, the left-sided MAPCAs were localized and readied for control. This often required dissection in the retroaortic area, in the area between the roof of the left atrium, the carina, and the left main-stem bronchus. The left side of the descending aorta was reached by then retracting the

ascending aorta rightward, continuing the dissection behind the superior aspect of the left atrium and sometimes around the esophagus. Opening the posterior pericardium was obviously required to reach these structures. This approach allowed exposure of MAPCAs arising from the lower descending aorta. Reaching left-sided collaterals arising from the upper descending aorta sometimes required dissection above the left main-stem bronchus. The native left pulmonary artery was mobilized at this point as well. Opening the left pleura was only necessary in the 13-year-old patient to repair a significant stenosis involving three branches of a large MAPCA with a pericardial patch. The right-sided pulmonary unifocalization was usually performed before cardiopulmonary bypass. We used the same guidelines that were previously described in the multiple-stage repair section to deal with the MAPCAs. Unlike Reddy, Liddicoat, and Hanley,⁷ in the presence of native pulmonary arteries, we ligated communicating collaterals, those supplying areas of the lung with dual blood supply and the small ones perfusing only one bronchopulmonary segment in any patient with a clearly adequate distal bed after unifocalization. We favored direct tissue-to-tissue anastomoses between native pulmonary arteries and MAPCAs and between collaterals. No prosthetic material was used in these patients. After the right-sided unifocalization, cardiopulmonary bypass was instituted with bicaval cannulation and immediately the remaining left-sided collaterals were controlled and ligated at their origins. Those targeted for unifocalization were then anastomosed accordingly either to the native left pulmonary artery or to other collaterals, while the patient was being cooled. Then the aorta was clamped and the intracardiac repair was performed as in group I. In one 10-week-old infant, the diminutive main pulmonary artery was used to establish right ventricular–pulmonary artery continuity with a transannular pericardial patch. In the other four patients the repair was effected with valved pulmonary homografts.

Follow-up. Before discharge, all patients had several echocardiograms to assess the adequacy of the repair. All survivors have been followed up by the referring cardiologist by clinical examination and echocardiograms. Cardiac catheterizations have been performed for suspected significant problems. Routine cardiac catheterizations are being planned in the future for the

Fig. 1. Pulmonary blood supply in patients undergoing the multiple-stage unifocalization followed by repair (group I).

Fig. 2. Pulmonary blood supply in patients undergoing single-stage midline unifocalization and repair (group II).

entire cohort. The status of all patients is known as of April 1997.

Results

Early results. Eleven of 12 patients (91.6%) underwent complete repair. In group I a mean of 3.3 operations were performed per patient to achieve repair; in group II, obviously, one operation per patient was done for repair. The preoperative diagnoses, operative details, surgical results, and outcome for each of the 11 patients to undergo complete repair are summarized in Tables III (group I) and IV (group II). One intraoperative death occurred in group I from unrecognized severe mitral stenosis discovered at autopsy. No early deaths occurred in group II. The right ventricular/left ventricular pressure ratio was 0.64 in the operating room, decreasing to 0.49 after the operation in group I as compared with 0.56 during the operation and 0.45 after the operation in group II (Fig. 3).

The complications for group I are listed in Table III and for group II in Table IV. Postoperative ventilatory support was required for a median duration of 6 days (range 1 to 233 days) in group I and of 9 days (range 8 to 23 days) in group II. In group I, two patients had severe bronchospasms. In one patient, these episodes resolved. In the other patient, the episodes of severe recurrent bronchospasms after the second unifocalization continued over a period of several months. Because of significant desaturations, the patient underwent complete repair and did well from a hemodynamic point of view with a right ventricular/left ventricular pressure ratio of 0.56. However, the episodes of bronchospasms continued. The patient required a tracheostomy and remained ventilator dependent until his death 7 months later of chronic lung disease. One patient had left phrenic nerve neurapraxia that resolved several months later. One patient had junctional ectopic tachycardia, which resolved. In group II, there were no episodes of severe bronchospasms or phrenic nerve problems. Two patients had transient vocal cord paralysis (one left and one right).

Table III. Clinical patient data: Multiple-stage unifocalization and repair

Patient	Age at repair	Diagnosis, preop anatomy	Previous operations	Final operation/repair	pRV/pLV at 24-48 hr	Complications
1	1.8 yr	TOF, PA, multiple VSDs, MAPCA $\times$ 2, discontinuous LPA	Continuity MPA-LPA (TtT); central shunt R unifocalization	Takedown shunt; VSD patch closure; RV-PA valved homograft	0.54	Severe bronchospasm early post/op
2	2.8 yr	TOF, PA, VSD, MAPCA $\times$ 3	L unifocalization (TtT) & L MBTS, R unifoc (Haz) & R MWS; revision bilateral central shunts	Takedown L and R shunts; VSD patch closure; RV-PA valved homograft	1.00	Operative death; autopsy; unrecognized severe mitral stenosis
3	9.3 yr	TOF, PA, VSD, MAPCA $\times$ 3	L unifocalization (TtT) and L MWS; R unifocalization (TtT) and R MWS	Takedown L and R shunts; VSD patch closure; RV-PA valved homograft	0.36	Junctional ectopic tachycardia
4	9.5 yr	TOF, PA, VSD, MAPCA $\times$ 6	Central shunt; L unifocalization (PTFE) and L MWS; R unifocalization and R MWS	Takedown L and R shunts; VSD patch closure; RV-PA valved homograft	0.39	None
5	3.3 yr	TOF, PA, VSD, MAPCA $\times$ 6, Di-George syndrome	L unifocalization and MBTS; coil embolization 1 L MAPCA; R unifocalization (PTFE) and R MWS	Takedown L and R shunts; VSD patch closure RV-PA valved homograft	0.56	Persistence of preop bronchospasm; ventilator dependence; late death from resp. comp. (7 mos postop)
6	3.8 yr	TOF, PA, VSD, MAPCA $\times$ 6	L unifocalization, MPA to ascending aorta; R unifocalization (PTFE) and L MWS	Takedown L and R shunts; VSD patch closure; RV-PA valved homograft	0.59	L phrenic nerve neurapraxia after L unifocalization and final repair; (full recovery)

TOF, Tetralogy of Fallot; PA, pulmonary atresia; VSD, ventricular septal defect; MAPCA, major aortopulmonary collateral artery; LPA, left pulmonary artery; MPA, main pulmonary artery; TtT, tissue-to-tissue anastomosis; R, right; L, left; MBTS, modified Blalock-Taussig shunt; Haz, hemiazygos vein tube graft; MWS, modified Watrston shunt; PTFE, polytetrafluoroethylene tube graft; RV-PA, right ventricle-pulmonary artery.

One patient had refractory junctional ectopic tachycardia necessitating extracorporeal life support for 3 days with full recovery.

Operative data and outcome for the two surgical approaches are compared in Table V. Repair was achieved at a much older median age of 3.5 years in group I as opposed to 28 weeks in group II. No significant differences were observed between the two groups in terms of circulatory arrest, aortic crossclamp time, or cardiopulmonary bypass time. One early and one late death occurred in group I and no early or late deaths in group II.

Follow-up. The families of all living patients were contacted in April 1997. The mean follow-up time after repair in group I was 23 months (range 1 to 72 months) and after complete repair in group II, 14 months (range 8 to 22 months). Four patients in group I and all five patients in group II are currently alive and well. The only reintervention in the two

groups was a successful percutaneous angioplasty of a left pulmonary artery stenosis 11 months after repair in a group II patient. The patient is currently alive and well.

Discussion

Pulmonary atresia, VSD, and MAPCAs is an extremely heterogeneous and complex cardiac malformation characterized by a highly variable pulmonary blood supply and arborization abnormalities. The native pulmonary arteries are either extremely diminutive (confluent or nonconfluent) or absent altogether. Pulmonary blood flow is provided by MAPCAs, which vary highly in number and origin. These are large and distinct arteries that usually arise from the descending thoracic aorta, but uncommonly may originate from the aortic arch or the subclavian, carotid, or even the coronary arteries. They may join end to end with an intrapulmonary

Table IV. Clinical patient data: Single-stage unifocalization and repair

Patient	Age at repair	Diagnosis, preop anatomy	Previous operations	Operation/repair	pRV/pLV at 24-48 hr	Complications
7	42 wk	TOF, PA, VSD, MAPCA × 3	None	Bilat unifoc (TtT); VSD patch closure; RV-PA valved homograft	0.50	Chylothorax on TPN × 3 wk
8	11 wk	DORV, PA, VSD, MAPCA × 3	None	Bilat unifoc (TtT); VSD enlargement with transannular patch; tunnel LV-Ao; RV-PA	0.55	Collapse and pneumothorax due to hypoplastic LLL
9	28 wk	d-TGA, CAVC, virtual PA, MAPCA × 4 L SVC, unroofed coronary sinus	None	Bilat unifoc (TtT); RV-PA valved homograft; patch repair of CAVC; enlargement with patch; tunnel LV-Ao; tunnel L SVC-RA; VSD	0.59	Refractory JET; cardiac arrest × 2; ECLS × 3 days (full recovery)
10	2 wk	TOF, PA, VSD, MAPCA × 4	None	Bilat unifoc (TtT); VSD patch closure; RV-PA valved homograft	0.29	Tamponade-chest opened × 7 days; JET; R vocal cord paralysis (full recovery)
11	13.7 yr	TOF, PA, VSD, MAPCA × 2	None	Bilat unifoc (BP & TtT); VSD patch closure; RV-PA valved homograft	0.30	Postop cardiac arrest-chest reopened × 2 days; L vocal cord paralysis (full recovery)

pRV/pLV, Right ventricular/left ventricular pressure ratio; TOF, Tetralogy of Fallot; PA, pulmonary atresia; VSD, ventricular septal defect; MAPCA, major aortopulmonary collateral artery; DORV, double-outlet right ventricle; d-TGA, D-transposition of the great arteries; CAVC, complete atrioventricular canal; L, left; SVC, superior vena cava; TtT, tissue-to-tissue anastomosis; RV-PA, right ventricle to pulmonary artery; LV-Ao, left ventricle to aorta; RA, right atrium; BP, bovine pericardium; TPN, total parenteral nutrition; LLL, left lower lobe; JET, junctional ectopic tachycardia; ECLS, extracorporeal life support; R, right.

artery, at which point the histologic appearance changes from that of a muscular artery to a pulmonary artery.⁹ Alternatively, they may connect end to side to a manifold of hilar pulmonary arteries or less commonly to a central pulmonary artery.^{10, 11} Therefore the MAPCAs are either communicating with the native pulmonary arteries or are not communicating.¹² They supply a variable territory of the lung, ranging from one or several segments, to one or several lobes, or even to an entire lung. An area of the lung may receive its blood supply from the native pulmonary arteries, from MAPCAs, or may have a dual blood supply. In the absence of stenosis, the MAPCAs may transmit systemic pressures to the pulmonary vascular bed, possibly leading to the development of pulmonary vascular obstructive disease in the area thus supplied. Often there is a varying degree of stenosis, protecting the distal pulmonary arterial bed.^{9, 13} These stenoses usually are the result of intimal proliferation and usually occur at branching points and at the junction of the collaterals with the native pulmonary arteries. Also, some of the collaterals may have a long and tortuous course, dampening the pressure transmitted to the distal vascular bed. By contrast, the intracardiac

component of the malformation is relatively straightforward, usually consisting of a large outlet VSD.

The natural history of these patients is complex and not easily determined because of the great variability of the structure of the pulmonary circulation. Because no two patients have exactly the same anatomy of the pulmonary circulation, available information is inadequate. The natural history is determined by the presence or absence of stenoses in the MAPCAs, with severe congestive heart failure at one extreme and severe hypoxia at the other. Without some intervention, most patients at these extremes die in early life. On the other hand, patients may have a relatively balanced pulmonary blood flow, either as a result of "optimal" stenoses or as a result of the development of pulmonary vascular obstructive disease in the areas supplied by "unprotected" MAPCAs. These patients remain relatively free of symptoms in early life, with the majority surviving until the teenage years. The cyanosis then begins to become worse, and most patients die by 30 years of age.¹⁴

Traditionally, the approach for these patients has been palliative, surgery being reserved only for the

Fig. 3. Comparison of postrepair right ventricular/left ventricular pressure ratio (pRV/pLV) in patients undergoing the multiple-stage versus the single-stage approach. *OR*, Operating room.

symptomatic patients with increased or decreased pulmonary blood flow. Several groups have advocated a more aggressive surgical approach, with an ultimate goal of complete biventricular repair through multiple pulmonary unifocalization procedures which, when successful, allow the intracardiac repair of the anomaly.¹⁻⁶ The final operation in the patients fulfilling the appropriate criteria is the intracardiac repair. This multiple-stage approach has resulted in complete repair in 11.5% to 60.5% of the patients in six large published series (Table VI).¹⁻⁶ The overall mortality to achieve complete repair has also been significant, ranging from 10.2% to 19.2%. Although this approach has not been universally successful, it has nevertheless offered hope in an otherwise hopeless situation. The downside is the requirement for multiple operations with no guarantee that a complete repair will be possible in each patient.

Recently, an aggressive new approach was reported in 10 patients by Reddy, Liddicoat, and

Hanley,⁷ consisting of a single-stage midline unifocalization and repair, with excellent results. There were no early deaths and one late death after a reoperation. Significant complications were phrenic nerve palsy in three patients and severe bronchospasm in three patients. This early experience suggests a significantly higher reparability rate than the traditional multistage approaches advocated by the other leading centers, while avoiding multiple operations. A summary of the experience with both approaches is found in Table VI.

At the Montreal Children's Hospital we have had experience with both these approaches in 12 patients. From 1989 to 1994, seven patients were subjected to the traditional multistage approach and complete repair was achieved in six. All five patients referred since 1995 have undergone the single-stage midline unifocalization and repair. With either approach, our intent was to intervene as early in life as possible, to take advantage of the possibly greater potential for growth of the respiratory system.¹⁵

Table V. Comparison of operative data and outcome between patients undergoing the multiple-stage and single stage repair

	Group I (n = 6): Multiple stage		Group II (n = 5): Single stage	
Age at repair in weeks: median and range	182	145-495	28	2-711
Number of operations: mean and range	3.3	3-4	1	1
pRV/pLV: mean and range				
At operation	0.64	0.40-1.00	0.56	0.40-0.73
24-48 hr	0.49	0.36-0.59	0.45	0.29-0.59
Operative times in minutes: mean and range				
Circulatory arrest	3	0-15	14	0-50
Crossclamp	130	120-139	143	69-197
Cardiopulmonary bypass	246	181-385	236	161-332
Days ventilated: median and range	6	4-233	9	8-23
Mortality (total)				
Early after repair		1		0
Late after repair		1		0
Follow-up in months: mean and range	23	1-72	14	8-22

pRV/pLV, Right ventricular/left ventricular pressure ratio.

Table VI. Summary of clinical series on repair of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries

First author (center)	Repair strategy	No. of patients	Median age (yr)	Overall mortality		Complete repairs	
				n	%	n	%
1988, Sullivan (Hospital for Sick Children, London, England)	Multiple stage	26	2.0	4	15.4	3	11.5
1989, Puga (Mayo Clinic, Rochester, Minn.)	Multiple stage	38	mean 4.7	4	10.5	23	60.5
1989, Sawatari (The Heart Institute, Tokyo, Japan)	Multiple stage	34	mean 6.6	4	11.8	10	29.4
1991, Iyer (Royal Children's Hospital, Melbourne, Australia)	Multiple stage	58	1.83	7	12.1	30	51.7
1994, Marelli (UCLA, Los Angeles, Calif.)	Multiple stage	26	mean 28	5	19.2	10	38.5
1996, Yagihara (National Cardiovascular Center, Osaka, Japan)	Multiple stage	50	mean 5.9	6	12.0	26	52.0
1995, Reddy (UCSF, San Francisco, Calif.)	Single stage	10	2.08	0	0	9	90.0
1997, Tchervenkov (Montreal Children's Hospital, Montreal, Canada)	Multiple and single stage (single-stage)	12 (5)	0.70 (0.54)	2 (0)	16.7 (0)	11 (5)	91.7 (100.0)

The mortality rate to achieve complete repair includes all deaths during preliminary operations, deaths between operations, and the early mortality after repair. It does not include late death after complete repair. UCLA, University of California; Los Angeles; UCSF, University of California, San Francisco.

Although all patients except one reached complete repair with the multistage approach, multiple operations were needed, and the patients' symptoms remained palliated, occasionally with lower saturation after the unifocalization. Furthermore, although the initial operation in the multistage approach was usually performed in infancy, the final repair was achieved only at a median age of 3.5 years. At the final repair, the posterior mediastinum and the hilar regions were significantly scarred,

increasing the surgical risk. The prolonged cyanosis and the previous operations appeared to stimulate the development of multiple secondary collaterals, potentially increasing the risk of bleeding. Finally, there was the risk of drop-off before the final repair, because of either interim death or the inadequate recruitment or growth of the distal pulmonary vascular bed. These reasons led us to adopt the single-stage approach. The desire to undertake full surgical repair in infancy, in addition to taking advantage of

the greater potential for growth of the respiratory system in early life, was also to avoid the deleterious effects of chronic cyanosis and volume overload of the heart and to prevent the development of pulmonary vascular obstructive disease in the unprotected collaterals. Although our experience with both approaches is still relatively small, we would like to make the following observations. All patients in the single-stage approach have undergone successful repair, with no early or late deaths. Repair was achieved at the initial operation at a median age of 28 weeks. Particularly encouraging is the low mean right ventricular/left ventricular pressure ratio of 0.45, which indicates excellent recruitment of an adequate pulmonary vascular bed. This is consistent with the results obtained by Reddy, Liddicoat, and Hanley,⁷ who achieved complete repair in 90% of their patients. For these reasons, we currently prefer the single-stage unifocalization and repair via a sternotomy. However, several limitations prevent us from making a firm and final conclusion. First, the follow-up is short and we do not yet have objective assessment of the long-term fate of the distal pulmonary arterial bed. The number of patients in each group is small, and it is uncertain whether the two groups are truly comparable. Finally, these are sequential series of patients rather than a prospective randomized study. However, because of the built-in biases of each surgical group and the uneven comfort level with infants and neonates, it is unlikely that a prospective randomized series will ever be done. Our unique experience with both approaches is unlikely to expand equally because, consistent with our overall philosophy, we are presently heavily biased toward the single-stage repair in infancy.

In conclusion, both the multistage unifocalization and single-stage approach can achieve complete biventricular repair in most patients if carefully planned and the operation accurately executed. However, the single-stage unifocalization and repair is our procedure of choice, because it can achieve a completely repaired heart and centralized pulmonary arteries with one operation in infancy.

REFERENCES

1. Sullivan ID, Wren C, Stark J, de Leval MR, Macartney FJ, Deanfield JE. Surgical unifocalization in pulmonary atresia and ventricular septal defect. A realistic goal? *Circulation* 1988;78(Suppl):III5-13.
2. Puga FJ, Leoni FE, Julsrud PR, Mair DD. Complete repair of pulmonary atresia, ventricular septal defect and severe peripheral arborization abnormalities of the central pulmonary arteries: experience with preliminary unifocalization procedures in 38 patients. *J Thorac Cardiovasc Surg* 1989;98:1018-29.
3. Sawatari K, Imai Y, Kurosawa H, Isomatsu Y, Momma K. Staged operation for pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries: new technique for complete unifocalization. *J Thorac Cardiovasc Surg* 1989;98:738-50.
4. Iyer KS, Mee RBB. Staged repair of pulmonary atresia with ventricular septal defect and major systemic to pulmonary artery collaterals. *Ann Thorac Surg* 1991;51:65-72.
5. Marelli AJ, Perloff JK, Child JS, Laks H. Pulmonary atresia with ventricular septal defect in adults. *Circulation* 1994;89:243-51.
6. Yagihara T, Yamamoto F, Nishigaki K, Matsuki O, Uemura H, Isizaka T, et al. Unifocalization for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. *J Thorac Cardiovasc Surg* 1996;112:392-402.
7. Reddy VM, Liddicoat JR, Hanley FL. Midline one-stage complete unifocalization and repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. *J Thorac Cardiovasc Surg* 1995;109:832-45.
8. Nakata S, Imai Y, Takanashi Y, et al. A new method for the quantitative standardization of cross-sectional areas of the congenital heart diseases with decreased pulmonary blood flow. *J Thorac Cardiovasc Surg* 1984;88:610-5.
9. Haworth SG, Macartney FJ. Growth and development of pulmonary circulation in pulmonary atresia with ventricular septal defect and major aorto-pulmonary collateral arteries. *Br Heart J* 1980;44:14-24.
10. Jefferson K, Rees S, Somerville J. Systemic arterial supply to the lungs in pulmonary atresia and its relation to pulmonary artery development. *Br Heart J* 1972;34:418-27.
11. Macartney FJ, Scott O, Deverall PB. Haemodynamic and anatomical characteristics of pulmonary blood supply in pulmonary atresia with ventricular septal defect—including a case of persistent fifth aortic arch. *Br Heart J* 1974;36:1049-60.
12. Faller K, Haworth SG, Taylor JFN, Macartney FJ. Duplicate sources of pulmonary blood supply in pulmonary atresia with ventricular septal defect. *Br Heart J* 1981;46:263-8.
13. McGoon MD, Fulton RE, Davis GD, Ritter DG, Neill CA, White RI Jr. Systemic collateral and pulmonary artery stenosis in patients with congenital pulmonary valve atresia and ventricular septal defect. *Circulation* 1977;56:473-9.
14. Kirklin JW, Barratt-Boyes BG. Tetralogy of Fallot with pulmonary atresia. In: Kirklin JW, Barratt-Boyes BG, editors. *Cardiac surgery*, 2nd ed. New York: Churchill Livingstone; 1993. p. 942-73.
15. Castaneda AR, Jonas RA, Mayer JE Jr, Hanley FL. Developmental biology: respiratory system. In: Castaneda AR, Jonas RA, Mayer JE Jr, Hanley FL, editors. *Cardiac surgery of the neonate and infant*. Philadelphia: WB Saunders; 1994. p. 18-22.

Discussion

Dr. Erle H. Austin III (Louisville, Ky.). The concept of early single-stage unifocalization and complete repair introduced by Frank Hanley is an important one that is being cautiously embraced by surgeons and cardiologists around the world. You are to be commended for your success with this approach and for providing one of the

first series to corroborate Dr. Hanley's excellent results. Comparing your recent experience with a prior personal series managed in a more conventional manner helps point out the potential advantages of single-stage complete repair in early infancy.

I thought that at least two technical details may have contributed to your good results. First, you were able to expose all of the major aortopulmonary collaterals by opening the posterior pericardium without entering the pleura. Second, in most cases you were able to complete the right-sided unifocalization before commencing bypass.

Although you have had success with this new approach, a careful review indicates that these are still difficult cases. Mean crossclamp time approached 2.5 hours and mean pump time approached 4 hours. Although all five patients survived, two of them experienced cardiac arrest after the operation, with one requiring 3 days of extracorporeal life support, and a third patient had tamponade necessitating emergency sternotomy. Surgeons contemplating the single-stage approach, therefore, should take these factors into consideration.

I have a couple of questions. First, your crossclamp times were longer than one would anticipate for VSD closure alone. In what ways did you use the additional crossclamp time to help with the reconstruction? Second, patients with major aortopulmonary collaterals are at risk of cerebral underperfusion during bypass. Did you take any special precautions with the patients in either group? Your follow-up indicated that all surviving patients were doing well hemodynamically. Did any patient in either group have a neurologic injury?

Dr. Tchervenkov. I agree with you that this is a novel procedure and it has to withstand the test of time. It is particularly exciting, though, that a full repair can be achieved in many of these patients with one operation. The extensive scarring and frozen mediastinum found at the final repair with the multiple-stage approach have made the single-stage repair a first choice for me.

I think my technique varies slightly from Dr. Hanley's technique in two ways. One is that we ligate the communicating collaterals. The second difference is that we expose the collaterals by opening the posterior pericardium, keeping in mind where the descending thoracic aorta is, usually without opening the pleura.

Why was the crossclamp time quite long? I believe that the most crucial part of the operation is the accurate reconstruction of the distal pulmonary arterial bed. This reconstruction is usually performed with the aortic clamp still on. I am not tempted to take the clamp off to shorten the ischemia time. I prefer to have an accurate anastomosis with a longer crossclamp time than the other way around. I have a similar experience with other complex repairs. At the last Society of Thoracic Surgeons meeting, I presented my experience with the arterial switch operation and concomitant aortic arch reconstruction in patients with transposition complexes with aortic arch obstruction with no operative mortality. Despite long crossclamp times and a single dose of crystalloid cardioplegic solution, these patients did well from a hemodynamic point of view.

In response to your question referring to the potential neurologic complications, the right-sided unifocalization

usually is accomplished before cardiopulmonary bypass at normothermia. The left-sided collaterals are localized and looped, ready to be snared as soon as bypass is instituted. I do not believe we have had any problems with hypoperfusion of the brain because all the collaterals are controlled the moment bypass is begun. We have not observed any significant neurologic complications, although one patient in the single-stage group had an absence of a corpus callosum with no change in status after the operation. Our series is still very small and it is still evolving. Unfortunately, I do not think it will expand equally between the multiple versus the single-stage approach because I am currently biased toward the single-stage approach.

Dr. Frank L. Hanley (San Francisco, Calif.). I would like to congratulate you for your excellent series. If evaluated from a historical perspective, this series is particularly important. As different approaches to this very complex lesion come into more focus, we find a number of groups who believe strongly in one particular approach. What is really needed is an objective perspective that examines different approaches. Although the current study is not a prospective, randomized, double-blind study, it is at least the first study that directly compares different philosophic approaches to this lesion. Ideally it will be a transition to a multicenter prospective trial of different approaches.

In my own personal commentary, a single-stage early repair only needs to be as successful as other repairs that necessitate multiple operations to be a superior approach. If there are any other potential benefits to the one-stage approach, addition to the reduced number of operations, the reduced number of hospitalizations, and the earlier age at which the repair occurs, then those add to the superiority of this approach.

Dr. Tchervenkov. Your comments are very pertinent. I agree with you that the surgeons who are set in their ways with either approach are probably unlikely to change. However, when the data from the various centers are carefully analyzed, it becomes apparent that in the two series with the single-stage approach, ours and yours, the mean or median age at recruitment is in infancy and is significantly lower than in the six large series that have been reported with the multistage approach. And I am not sure we are talking about exactly the same patients. In some of these centers, many of the patients are distant referrals and are in a way preselected. It is very hard to objectively compare the two approaches. I think that a prospective randomized study is relatively unlikely, because of the uneven experience among centers with complex neonatal and infant repairs, so we have to look at how things evolve over time. Of particular importance is the fate of the distal pulmonary arterial bed. Are a lot of these anastomoses going to perhaps narrow down and then require further intervention, such as dilatation and stenting? This question, however, applies equally to both the single-stage and the multiple-stage approaches. I think we are just at the beginning.

Dr. Nitu V. Mandke (Mumbai, India). I congratulate you on the excellent results. Of course a series of this kind is going to be small. I would like to share some of our experiences and offer a suggestion.

In the past 2 years we have performed one-stage unifocalization and total correction via a median sternotomy in three patients. We found it very easy to transect the ascending aorta and retract it to provide easy access to the MAPCAs and then do the operation. I think it is easier and much more comfortable to do this kind of procedure with all the MAPCAs on both the sides of the descending thoracic aorta. We found it an extraordinary approach. One can really get good vision and good anastomosis and then one can rejoin the aorta and then do the conduit. Of course, the conduit can be anastomosed first after the unifocalization and then the ascending aorta can be reconnected.

Dr. Tchervenkov. This is an interesting comment. I do not hesitate to transect the ascending aorta if I have to access centrally hypoplastic, stenotic native pulmonary arteries to enlarge them and not to have to work behind the aorta if it is awkward. However, transecting the aorta to expose the MAPCAs implies that bypass is needed, and that increases the ischemia time. Furthermore, as Dr. Austin alluded to, starting bypass before all the MAPCAs are adequately controlled may increase the potential for neurologic complications. I am not sure that I would personally accept your suggestion.

Dr. Hanley (*San Francisco, Calif.*). I would like to make one final comment about the neurologic issue. Many of the studies that look at these patients emphasize the completeness of the repair and the hemodynamic results. Many of the large studies that perform staged repairs with cardiopulmonary bypass without a complete repair and without control of the collaterals have shown an

extraordinarily high incidence of neurologic injury, including choreoathetosis, sometimes in as many as 25% or 30% of these patients. One of the greatest advantages of the one-stage repair, as Dr. Tchervenkov has emphasized, is that one gains control of all collaterals before starting bypass, usually within several minutes of the start of bypass. All of these collaterals can be controlled and there is no further runoff. The bypass run is really no different from that used in a VSD operation in terms of potential neurologic injury. The bypass time may be longer, but the physiology of bypass is such that potential runoff into the lungs is essentially nonexistent. In our series, now with 59 patients, other than one patient who had a stormy postoperative course, required extracorporeal membrane oxygenation, and did have a neurologic insult from bleeding during extracorporeal membrane oxygenation, there has not been any neurologic injury.

Dr. Tchervenkov. This is an important point. When the single-stage approach is begun in early life, the development of the secondary collaterals that are particularly bothersome in the older patients is avoided. You can control the major collaterals ahead of time before bypass. However, if you are operating on an older patient who has multiple secondary collaterals in the mediastinum, the runoff in those vessels cannot be controlled. The single-stage approach offers many advantages and I hope that increasing experience will lead to better surgical techniques, improved results, and the earlier referral of patients before the long-term complications of the unrepaired malformation set in.