Staged repair of tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries

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Objective: To assess the results of a staged surgical approach for tetralogy of Fallot with pulmonary atresia, hypoplastic or absent pulmonary arteries, and major aortopulmonary collateral arteries.

Methods: We retrospectively reviewed a consecutive series of these patients from a single institution.

Results: From July 1993 to April 2001, 46 consecutive patients with tetralogy of Fallot, pulmonary atresia, and major aortopulmonary collateral arteries were treated with staged surgical repair. The operative sequence usually began with a central aortopulmonary shunt followed by unifocalization of aortopulmonary collateral arteries depending on the source and distribution of pulmonary blood flow. Twenty-eight patients (61%) subsequently underwent complete repair with ventricular septal defect closure and right ventricle to pulmonary artery connection. Those patients who underwent complete repair had a median of 3 total operations (range 1-6). The ratio of the mean pulmonary artery pressure to the mean systemic blood pressure at the time of complete repair was 0.36 (range 0.19-0.58). Two of the 28 repaired patients (7.1%) required subsequent fenestration of the ventricular septal defect closure due to later development of supersystemic right ventricular pressure and right ventricular failure. Eighteen patients (39%) have undergone 1 or more staging operations and are considered good candidates for eventual complete repair. There were no hospital deaths. There was 1 late death (2.2%; 95% CI 0.4-11.3%) in a patient born prematurely who developed severe bronchopulmonary dysplasia precluding complete repair.

Conclusions: For tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries, a staged surgical approach yields low overall mortality and acceptable hemodynamics after complete repair.

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Received for publication Dec 2, 2002; revisions requested March 10, 2003; revisions received March 25, 2003; accepted for publication April 11, 2003.

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J Thorac Cardiovasc Surg 2003;126:694-702

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0022-5223/2003 \$30.00 + 0

doi:10.1016/S0022-5223(03)00700-1

Treatment goals for patients who have tetralogy of Fallot with pulmonary atresia (TOF-PA) and major aortopulmonary collateral arteries (MAPCAs) are aimed at establishing a central source of pulmonary blood flow sufficient to allow closure of the ventricular septal defect (VSD) with low and stable right ventricular pressure.¹⁻¹⁴ A staged approach to this problem, developed at the Royal Children's Hospital in Melbourne, employs an initial central shunt to hypoplastic central pulmonary arteries, if present, to encourage the growth and development of the native pulmonary arterial system.¹ Subsequent procedures incorporate pulmonary blood flow from MAPCAs into a single source based on the native pulmonary arteries (unifocalization). When central pulmonary arteries are absent, branch pulmonary arteries are created into which MAPCAs are transplanted. After unifocalization, the VSD is closed and right ventricle to pulmonary artery continuity is established as the final step in this sequence. The following constitutes a description of this staged approach utilized at The Children's Hospital at The Cleveland Clinic.

Methods

Patients

From July 1993 to April 2001, 46 consecutive patients with TOF-PA and MAPCAs were treated with staged surgical repair (Table 1). No patient died awaiting surgery. Fifteen patients had 1 or more initial operative procedures (19 total) performed at other institutions (Table 2), and the remainder were seen primarily at this institution.

Operative Approach: Central Shunt

All patients had delineation of the native pulmonary artery and MAPCA anatomy made by cardiac catheterization prior to each stage of the operative sequence (Tables 3 and 4). Central pulmonary artery anatomy was established by direct injection of a shunt, if present, or by retrograde filling after MAPCA or pulmonary venous wedge injection. Six patients (15%) had absence of the central pulmonary arteries, and the remainder had diminutive central pulmonary arteries. The operative approach for patients with diminutive but confluent central pulmonary arteries was determined by the physiologic status of the patient (Figure 1).¹ The first step in the sequence was to create a central shunt from the aorta to the transected main pulmonary artery (Melbourne shunt).¹⁵ If a Melbourne shunt was performed as an isolated procedure, a median sternotomy was used; however, if the central pulmonary arteries were larger than diminutive (as an approximation, >2.5 mm diameter in an infant) and if concurrent left-sided unifocalization of MAPCAs was considered feasible, this was performed through a left thoracotomy (Figure 2). The pulmonary arteries were controlled with soft clamps. The main pulmonary artery was transected from the right ventricular outflow tract, and the opened end of the main pulmonary artery was filleted and sewn directly to the left posterior surface of the ascending aorta to avoid tension on the right pulmonary artery (Figure 3).

TABLE 1. Demographics for patients with TOF-PA and MAPCAs at presentation for first operation

Age (median)	7.2 months (range 17 days to 22.8 years)
Weight (median)	6.6 kg (range 3.0-50.0 kg)
Microdeletion of 22q11	7 patients (15%)
Prior operations	15 patients (33%)
Oxygen saturation (median)	74% (range 50-90%)

TABLE 2. Previous operations performed on 15 patients at outside institutions

Melbourne shunt
Left unifocalization
Right ventricular outflow tract patch, bilateral unifocalization
(1) Central shunt (2) RV-PA conduit, central shunt to right MAPCA
Right thoracotomy, right MBTS, ligation of 2 MAPCAs
RV-PA conduit, right unifocalization
Right MBTS, ligation of MAPCA
Right unifocalization, right MBTS
Left unifocalization, left MBTS
Bilateral unifocalization, bilateral MBTS
(1) Right unifocalization (2) left unifocalization, left MBTS
(1) Right MBTS (2) left unifocalization, left MBTS
Left thoracotomy, ligation of 2 MAPCAs
(1) Right unifocalization (2) left unifocalization
Right ventricular outflow tract patch

RV-PA, Right ventricle to pulmonary artery; MAPCA, major aortopulmonary collateral artery; MBTS, modified Blalock-Taussig shunt. Four patients had 2 prior operations, listed in parentheses as (1) and (2).

TABLE 3. Pulmonary artery and MAPCA anatomy

Pulmonary artery anatomy	Number
Patients with hypoplastic pulmonary arteries	40 (85%)
Patients with absent central pulmonary arteries	6 (15%)
Total number of MAPCAs	133
Median number of MAPCAs/patient	3 (range 1-6)

TABLE 4. Number and location of MAPCA origins

	Number (%)
Descending aorta	104 (78.2)
Aortic arch	7 (5.3)
Left subclavian artery	6 (4.5)
Ascending aorta	5 (3.8)
Right subclavian artery	4 (3.0)
Left internal thoracic artery	1 (0.8)
Right coronary artery	1 (0.8)
Left common carotid artery	1 (0.8)
Unspecified	4 (3.0)
Total	133

Figure 1. Paradigm for selective, staged treatment of TOF-PA with diminutive confluent central pulmonary arteries.

Figure 2. Paradigm for selective, staged treatment of TOF-PA with small confluent central pulmonary arteries.

Operative Approach: Unifocalization and Ligation of MAPCAs

All large MAPCAs were considered suitable for unifocalization even if there was overlap with native pulmonary arterial distribution or if there were small communications with native pulmonary arteries. MAPCAs were ligated if they were small and supplied an area of the lung judged to be less than a single bronchopulmonary segment.

One hundred thirty-three MAPCAs were present in these 46 patients; 18 MAPCAs were approached operatively prior to treatment at this institution. Of the 115 MAPCAs managed operatively at this institution, 39 (34%) were ligated and 76 (66%) were unifocalized. Patients who had congestive heart failure underwent unifocalization initially on the side that provided the most unobstructed pulmonary blood flow. Patients with significant cyanosis had unifocalization performed initially on the side with the most obstructed MAPCAs. In either case, the contralateral side was unifocalized within weeks to months after the initial unifocalization. For unifocalization, MAPCAs were soft clamped and then detached from their systemic blood supply. The transected end was then beveled and sewn end-to-side to the native pulmonary artery using 7-0 polypropylene suture.¹ The use of foreign material to unifocalize MAPCAs was avoided in all cases. For MAPCAs that had to be mobilized for some distance to allow anastomosis to the central pulmonary arteries (usually from the distal descending aorta), an interposition graft of reversed, native azygos vein was used when possible.¹⁶

Operative Approach: Completion of Repair

Patients were considered for completion of the repair with VSD closure when (1) no sizable MAPCAs remained, (2) more than approximately two thirds of the bronchopulmonary segments were connected to the central pulmonary blood supply, and (3) the estimated resistance in the distal pulmonary arterial bed was judged to be low. Repair was performed through a median sternotomy using cardiopulmonary bypass as described.¹ All sources of pulmonary blood flow were controlled with the institution of cardiopulmonary bypass. With the heart beating, all stenoses of the central pulmonary arteries were reconstructed with fresh autologous pericardium. After aortic crossclamping, a vertical right ventriculotomy was performed, potentially obstructive muscle bundles were resected, and the VSD was closed with a knitted Dacron patch in all cases. The aortic crossclamp was then removed, and the right ventricular outflow tract was reconstructed with either a Hancock valved conduit or a pulmonary allograft. A fine pressure line (Intracath, 22 gauge $\times$ 8 inch; Becton Dickinson Vascular Access, Sandy, Utah) was placed into the pulmonary artery for determination of the mean pulmonary arterial pressure at the completion of the repair.

Operative Approach: TOF-PA with Absent Central Pulmonary Arteries

As for patients with confluent pulmonary arteries, the operative approach was determined by the physiologic status of the patient (Figure 4). Sides were approached sequentially by creating a roll of autologous pericardium (preferably) or remodeled pulmonary al-

Figure 3. Melbourne shunt: side-biting clamp controls the ascending aorta; soft clamps control the branch pulmonary arteries. *Inset* demonstrates the completed shunt with the pulmonary artery anastomosed to the posterior and left lateral aspect of the ascending aorta close to the sinotubular junction. Reprinted with the permission of The Cleveland Clinic Foundation.

Figure 4. Paradigm for selective, staged treatment of TOF-PA with absent central pulmonary arteries.

lograft.^{1,17,18} MAPCAs were then anastomosed to these neopulmonary arteries, which were brought into the mediastinum via pericardial windows behind the phrenic nerves and tacked to the ascending aorta. A polytetrafluoroethylene shunt derived from the ipsilateral subclavian artery was then anastomosed to the neopulmonary artery to provide its systemic arterial supply. Subsequently, via median sternotomy, the anatomically placed neopulmonary arteries were connected behind the ascending aorta and joined to the right ventricle by valved conduit after VSD closure.

Statistical Analysis

Student paired *t* test was used to compare the intraoperative mean pulmonary artery to mean systemic pressure ratio to the latest determination of right ventricular systolic pressure to systemic systolic blood pressure ratio using JMP Statistical Discovery Software (SAS Institute, Cary, NC). This analysis was repeated omit-

ting the 2 patients who developed systemic right-sided pressures necessitating VSD patch fenestration (see Results below).

Results

One hundred-nine operative procedures were performed in these 46 patients (median 3 operations per patient; range 1-6 per patient). Of the 31 patients who had their initial surgery performed at this institution, 26 had a Melbourne shunt with or without concomitant unifocalization (Table 5). For patients who underwent an initial Melbourne shunt, the mean pulmonary artery pressure was low in most cases at subsequent cardiac catheterization (median mean pulmonary artery pressure 24.5, range 12-62). Three of the 31 patients underwent all unifocalization via a single thoracotomy as their initial procedure. Two of the 31 patients had absent

TABLE 5. Initial surgical procedures performed in 46 patients

Procedure	Patients treated primarily	Patients with prior operations
Melbourne shunt	20	1
Melbourne shunt $\pm$ MAPCA unifocalization/ligation	6	
Repair (VSD closure, RV-PA connection)		5
Left MAPCA unifocalization/ligation $\pm$ shunt	2	6
Right MAPCA unifocalization/ligation $\pm$ shunt	1	3
Creation of neopulmonary artery with shunt	2	
Total	31	15

MAPCA, Major aortopulmonary collateral artery; VSD, ventricular septal defect; RV-PA, right ventricle to pulmonary artery.

central pulmonary arteries and underwent creation of a unilateral neopulmonary artery for subsequent creation of a central pulmonary arterial confluence as their initial procedure. Fifteen patients had prior operations at other institutions, 5 of whom were able to have their repair completed by VSD closure and establishment of right ventricle to pulmonary artery continuity as the initial procedure at this institution. Of the remaining 10 patients who had undergone prior procedures elsewhere, 1 patient received a Melbourne shunt and the remainder had some type of unifocalization as their initial procedure, including 4 patients who initially had absent central pulmonary arteries.

Sixty-three operations were performed in the remaining 41 patients who had not undergone complete repair during the initial operation (Table 6). Twenty-three of these 63 operations were complete repairs with VSD closure and establishment of right ventricle to pulmonary artery continuity.

Twenty-eight of the 46 patients (61%) have undergone complete repair with VSD closure and establishment of right ventricle to pulmonary artery continuity at a median age of 2.9 years (range 5.7 months to 22.8 years). The median interval between the first operation and complete repair for the 23 patients in whom their first operation at this institution was not complete repair was 17.6 months (range 6.5-45.9). Including procedures performed at other institutions, these 28 patients required a median of three operations to achieve complete repair (Figure 5). The median cardiopulmonary bypass time for these 28 patients at the time of complete repair was 163 minutes (range 124-287) with a median crossclamp time of 36.5 minutes (range 22-70). At the completion of the repair the mean pulmonary

TABLE 6. Subsequent surgical procedures performed in 41 patients who had not undergone complete repair as their initial surgical procedure

Procedure	Number
Repair (VSD closure, RV-PA connection)	23
Left MAPCA unifocalization/ligation $\pm$ shunt	13
Right MAPCA unifocalization/ligation $\pm$ shunt	18
Shunt $\pm$ PA'plasty	5
Other	4
Total	63

VSD, Ventricular septal defect; RV-PA, right ventricle to pulmonary artery; MAPCA, major aortopulmonary collateral artery; PA'plasty, pulmonary arterioplasty.

artery pressure to systemic arterial pressure ratio determined in the operating room had a median value of 0.36 (range 0.19-0.58). For the complete repair, the median duration of intubation for these 28 operations was 1 day (range 0-34); the median duration of intensive care unit stay was 2 days (range 1-38); and the median duration of total hospital stay was 7 days (range 5-44).

Complications

There were 31 complications in the 109 procedures (28%) in 20 patients (Table 7). There were 3 cases of cardiac arrest: 1 occurred due to cardiac perforation during pericardiocentesis; a second cardiac arrest occurred due to acute respiratory failure after extubation; the etiology of the third cardiac arrest was unexplained. In each of the 3 cases the duration of cardiac arrest was brief and recovery was complete.

Outcome

Figure 6 demonstrates the outcomes of these 46 patients. There were no hospital deaths. Two of the 28 patients who underwent complete repair developed progressive increases in right ventricular pressure, without new development of peripheral pulmonary artery stenoses, necessitating operative fenestration of the VSD patch 2 and 3 months postoperatively. A single patient who had 3 previous staging procedures but in whom VSD closure was not an option due to high pulmonary artery resistance ultimately died months after the last operative procedure from bronchopulmonary dysplasia (mortality rate 2.2%; 95% CI 0.4-11.3%). The remaining 17 patients have undergone 1 or more staging procedures and remain in the sequence as candidates for ultimate VSD closure.

Long-term Follow-up After Complete Repair

Follow-up was available for 26 of the 28 patients (93%) who underwent complete repair, including the 2 patients with subsequent VSD fenestration (median follow-up 44

TABLE 7. Complications

Complication	Number
Occlusion of shunt or reconstructed PA requiring urgent reoperation	3
Thoracentesis	3
Sepsis	3
Pericardial effusion requiring pericardiocentesis	3
Respiratory failure requiring prolonged ventilation	3
Chronic occlusion of shunt or reconstructed PA	2
Diaphragmatic paralysis	2
Hypoxemia requiring emergency shunt	2
Cardiac arrest at time of failed extubation	1
Cardiac arrest due to cardiac perforation at the time of pericardiocentesis	1
Cardiac arrest (unexplained)	1
Necrotizing enterocolitis	1
Left cerebral cortical infarction	1
Excess MAPCA flow requiring unifocalization	1
Aneurysmal dilatation of pericardial roll used for PA reconstruction	1
Supraventricular tachycardia	1
Thrombectomy of femoral artery thrombosis	1
Reexploration for bleeding with tamponade	1
Total	31

PA, Pulmonary artery; MAPCA, major aortopulmonary collateral artery.

months, range 1-79). The right ventricular to left ventricular pressure ratio during the period of follow-up was significantly higher than the intraoperative mean pulmonary artery to mean systemic pressure ratio (0.50 ± 0.22 versus 0.35 ± 0.10 ; $P = .01$; Figure 7). The latest right ventricular pressures were either measured directly at the time of cardiac catheterization or estimated by echocardiography. Excluding the 2 patients who developed systemic right ventricular pressure necessitating VSD fenestration still resulted in significantly higher right-sided pressures during follow-up (0.45 ± 0.16 versus 0.35 ± 0.10 ; $P = .04$). Twenty interventions performed in either the cardiac catheterization laboratory (14 procedures) or the operating room (6 procedures) were required in 14 of the 26 patients (54%) for whom follow-up is available after complete repair; 3 of these patients required multiple interventions (Table 8).

Discussion

The anatomic goals of treatment for patients with TOF-PA and MAPCAs are to provide a partitioned circulation by VSD closure and to establish continuity between the right ventricle and the pulmonary arteries. Of equal importance to these anatomic goals, however, is the establishment of a physiologic status for the pulmonary circulation that leads to low and stable right ventricular pressure by recruiting all

Figure 5. Number of operative procedures for patients ultimately undergoing complete repair.**TABLE 8. Interventions after complete repair**

Procedure	Number
Balloon dilatation of pulmonary arteries	12
Balloon dilatation of conduit stenosis and pulmonary arteries	1
Balloon dilatation of pulmonary arteries and coiling of MAPCAs	1
Replacement of RV-PA conduit	3
VSD fenestration	2
Repair of RVOT aneurysm	1
Total	20

MAPCAs, Major aortopulmonary collateral arteries; RV-PA, right ventricle to pulmonary artery; VSD, ventricular septal defect; RVOT, right ventricular outflow tract.

available sources of pulmonary blood flow. A staged approach to this condition enhances the opportunity for central pulmonary artery development, provides maximal runoff for the pulmonary circulation by extensive unifocalization, and optimizes the chances for survival. We have found that this staged approach best provides individualized treatment for these children by matching the operative approach to the specific anatomy and physiology of each patient.

The initial performance of a central shunt has reliably provided impetus for uniform growth and development of the central pulmonary arteries. The Melbourne shunt utilizes only native tissue to provide a centralized source of pulmonary blood flow and can be performed without cardiopulmonary bypass in almost all cases.¹⁵ Early after the Melbourne shunt, the aortic pressure is conducted to the small pulmonary arteries without obstruction. As pulmonary arterial growth occurs, pulmonary blood flow increases exponentially, which can lead to congestive heart failure or the development of pulmonary vascular occlusive disease. In most patients, however, the orifice of the shunt remains small compared with the increasing size of the branch pulmonary arteries; in these cases, pressures become low

Figure 6. Outcomes for patients with TOF-PA treated with selective, staged surgical protocol.

Figure 7. Intraoperative mean pulmonary artery pressure to mean systemic blood pressure ratios (MPAP/MSBP) and latest right ventricular pressure to systemic systolic blood pressure ratios (RVP/SBP) for patients undergoing complete repair (both ratios available for 22 patients). Filled diamonds, patients who remain with closed VSD; clear diamonds, patients who have undergone VSD fenestration after VSD closure (n = 2).

and the risk of early pulmonary vascular occlusive disease appears to be obviated. As MAPCAs are added, runoff increases, resulting in a further decrease in pulmonary artery pressure. With these points in mind, patients require close follow-up after the Melbourne shunt and unifocalization followed by complete repair as soon as satisfactory pulmonary artery growth has occurred.

The timing of entry into the operative sequence is important; in our experience younger patients are most likely to respond with growth of the native pulmonary arteries sufficient to allow full repair. Later referral of patients may result in a missed opportunity for maximal native pulmonary artery development after central shunting. Poor outcomes in patients referred late for surgical therapy may also reflect an unfavorable selection bias. Patients who possess high flow through MAPCAs may be referred late due to the

absence of clinically significant cyanosis. These patients are at risk for the development of obstructive vascular disease in pulmonary segments supplied by unobstructed MAPCAs.^{19,20} Older patients without high MAPCA flow due to proximal stenoses have diminished risk for some of these problems, however; in our experience, these patients are more likely to develop profuse small mediastinal collaterals (not MAPCAs) that make operations tedious and difficult. We continue to recommend that patients enter the operative sequence by 6 months of age for elective intervention but patients who have profound cyanosis or congestive heart failure should enter earlier.

Interval performance of unifocalization after central shunting enhances the prospects of anastomosing important MAPCAs to an adequately developed central pulmonary arterial system, leading to maximal recruitment of all

TABLE 9. Published reports and the present series for treatment of TOF-PA

Institution	Number of patients	Hospital survival (%)	Intraoperative RV/LV pressure	Long-term survival (%)	Latest RV/LV pressure	Length of follow up (months)
Royal Children's Hospital, Melbourne, 1991 ¹	58	97	0.51	90	NR	43
Children's Hospital, Boston, 1993 ⁶	48	73	NR	56	0.71	120
University of California, Los Angeles, 1994 ¹²	12	100	0.45	100	NR	9
University of Michigan, 1995 ²⁶	14	79	0.57	64	NR	9
Children's Hospital, Los Angeles, 1997 ¹³	10	100	NR	100	0.44	17
Montreal Children's Hospital, 1997 ⁸	12	92	0.47	83	NR	19
Bambino Gesù, Rome, 1998 ¹⁰	15	93	0.48	93	0.42	14
Children's Mercy Hospital, Kansas City, 2000 ²⁷	11	91	<0.5	91	NR	NR
University of California, San Francisco, 2000 ⁵	85	89	0.44	81	NR	22
Children's Hospital La Timone, Marseille, 2001 ¹¹	10	90	0.5	80	0.6	45
Cleveland Clinic Children's Hospital	46	100	0.36	98	0.50	44

TOF-PA, Tetralogy of Fallot and pulmonary atresia; RV/LV, right ventricular/left ventricular; NR, not reported.

sources of pulmonary blood flow, which may contribute to gas exchange and runoff for the pulmonary circulation. It has been demonstrated that MAPCAs branch within the lung parenchyma into progressively smaller units that terminate in efficient respiratory units, similar to native pulmonary arteries.¹⁹⁻²² Staged unifocalization allows an individualized approach to be made in each case by optimally matching the most appropriate operative procedure to the existing anatomy and physiology. For patients who have congestive heart failure, unifocalization is initially performed on the side with the least obstructed pulmonary blood flow, which makes congestive heart failure easier to manage and decreases the likelihood of development of obstructive disease in overcirculated pulmonary segments. Patients with significant cyanosis have unifocalization performed initially on the side with the most obstructed MAPCAs. Modified Blalock-Taussig shunts are often performed adjunctively at the time of unifocalization in these cases to further augment pulmonary blood flow with a resulting decrease in cyanosis. Staged unifocalization may not always require bilateral thoracotomies in addition to median sternotomy. Large central MAPCAs that originate relatively close to the central pulmonary arteries (especially those on the left side) may be easily unifocalized at the time of complete repair through a median sternotomy. In the majority of cases all unifocalization and ultimate complete repair can be performed within a year after entering the operative sequence that commences with the performance of a central shunt.

The operative approach described herein reliably provides centralized pulmonary blood flow to more than two thirds of the total bronchopulmonary segments, which we have established as the necessary outflow for successful VSD closure. We have not found measures of central pulmonary artery size, such as the Nakata index²³ or the McGoon ratio,²⁴ to be helpful in determining which patients are suitable for repair because of their inability to predict the

anatomy and physiologic status of the peripheral pulmonary circulation. In this series, more than 60% of the patients have completed repair and only 2 patients (7%) who underwent VSD closure subsequently required fenestration of the VSD patch due to the development of supersystemic right ventricular pressures. Interestingly, the intraoperative mean pulmonary artery to systemic arterial pressure ratios were low (0.36 and 0.43) in these 2 patients. The development of supersystemic right ventricular pressures occurred months after repair due to progressive stenoses of small pulmonary vessels distal to what could be approached in the cardiac catheterization laboratory. All of the patients who remain in the operative sequence are considered to be acceptable candidates for complete repair.

During the period of follow-up, right ventricular to systemic pressure ratios increased compared with intraoperative measurements even after excluding the 2 patients who required VSD takedown. Although the true pulmonary arterial pressure may have been underestimated in the immediate postoperative period due to the small-gauge catheter used in these patients, this increase in pulmonary arterial pressure was seen in 11 of 22 patients (Figure 7). This finding may suggest that the distal pulmonary vasculature is abnormal and prone to progressive occlusive disease. An additional contributing factor may be preferential blood flow to lung segments with the lowest resistance, predisposing these areas to occlusive vascular disease, even in patients with "acceptable" postrepair pulmonary artery pressures (<60 mm Hg).

Due to the complex nature of this disease a number of operative approaches have evolved for its treatment (Table 9). Reddy and coworkers⁵ described their experience with single-stage unifocalization and full intracardiac repair. Sixty-six percent of these patients were able to have single-stage unifocalization and full intracardiac repair, and the remainder underwent staging of unifocalization and repair. Four patients required early reoperation for incorrect deci-

sions about VSD closure; however, after adoption of an intraoperative pulmonary blood flow study, no other patients required early reoperation for VSD closure or take-down.²⁵ Early mortality was 10.6%, which was statistically associated with duration of cardiopulmonary bypass, and overall mortality was 18.8%.

In summary, for patients with TOF-PA we advocate a flexible surgical program that matches the operative procedure to the individual patient's anatomy and physiology. Staged unifocalization establishes controlled pulmonary blood flow, which avoids exposing unprotected lung segments to systemic pressure prior to VSD closure.

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