Early and Intermediate Outcomes After Repair of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries
Experience With 85 Patients

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Background—Pulmonary atresia with ventricular septal defect (VSD) and major aortopulmonary collaterals (MAPCAs) is a complex lesion with marked heterogeneity of pulmonary blood supply. Traditional management has involved staged unifocalization of pulmonary blood supply. Our approach has been to perform early 1-stage complete unifocalization in almost all patients.

Methods and Results—Since 1992, 85 patients with pulmonary atresia, VSD, and MAPCAs have undergone unifocalization (median age, 7 months). Complete 1-stage unifocalization and intracardiac repair were performed through a midline approach in 56 patients, whereas 23 underwent unifocalization in a single stage with the VSD left open, and 6 underwent staged unifocalization through sequential thoracotomies. There were 9 early deaths. During follow-up (1 to 69 months), there were 7 late deaths. Actuarial survival was 80% at 3 years. Among early survivors, actuarial survival with complete repair was 88% at 2 years. Reintervention on the neo–pulmonary arteries was performed in 24 patients.

Conclusions—Early 1-stage complete unifocalization can be performed in >90% of patients with pulmonary atresia and MAPCAs, even those with absent true pulmonary arteries, and yields good functional results. Complete repair during the same operation is achieved in two thirds of patients. There remains room for improvement; actuarial survival 3 years after surgery is 80%, and there is a significant rate of reintervention. These results must be appreciated within the context of the natural history of this lesion: 65% of patients survive to 1 year of age and slightly >50% survive to 2 years even with surgical intervention. (*Circulation*. 2000;101:1826-1832.)

Key words: survival ■ surgery ■ heart defects, congenital

Pulmonary atresia with ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs) is a complex congenital anomaly characterized by heterogeneous and frequently severe anomalies of pulmonary blood supply. The complexity of this lesion is such that, until the mid-1970s, most patients were managed medically, with surgical palliation, or with right ventricular (RV) outflow reconstruction and ligation of MAPCAs.[1–3] Since the detailed characterization of MAPCAs,[4–9] several investigators have developed programs to manage this anomaly. These have typically incorporated a strategy of staged unifocalization, with the initial operation designed to increase flow to the true pulmonary arteries (PAs) in an effort to stimulate growth.[10–20] Although these various strategies have contributed to our understanding of this lesion and have provided satisfactory results for a select group of patients, most leave a substantial proportion of patients without complete repair.

In an attempt to improve the prospects for patients with pulmonary atresia, VSD, and MAPCAs, we initiated in 1992 a program of 1-stage unifocalization and repair for this lesion. In our initial report, we described our experience with 11 patients managed with this approach.[21] Seventy-four additional patients with pulmonary atresia and MAPCAs have since undergone surgery at our institution. We present our experience with these 85 patients.

Methods

Patients

From July 1992 through April 1998, 89 patients with pulmonary atresia, VSD, and MAPCAs were referred for surgery. Of these, 4 patients (3 infants), all of whom had been referred from out of state, died before the scheduled date of surgery. No referred patients were refused as surgical candidates. The remaining 85 patients underwent surgery and constitute the study group for this report. Age at surgery

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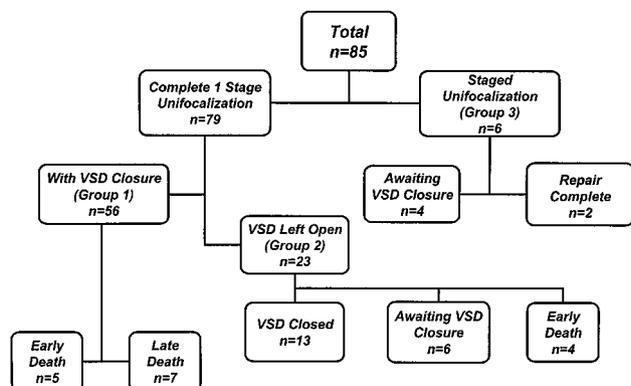
Figure 1. Flow diagram demonstrating treatment method and outcomes in 85 patients with pulmonary atresia, VSD, and MAPCAs managed at our institution between July 1992 and April 1998.

ranged from 10 days to 37 years (median, 7 months), and 57 patients (67%) were <1 year of age. Previous operations had been performed in 14 patients, including systemic-to-PA shunts (n=11), partial unifocalization (n=2), and banding of a large collateral (n=1). All these surgeries were performed at other institutions, except for the collateral banding, which we performed in a neonate who developed necrotizing enterocolitis as a result of severe pulmonary overcirculation. The total number of MAPCAs was 322, with a median of 4.[1–7] Almost 90% arose from the descending aorta, whereas 34 originated from the brachiocephalic vessels or their branches (n=14), the aortic arch (n=4), or the coronary arteries (n=6). Fourteen patients had absent true PAs as determined by intraoperative inspection. Twelve patients (14%) were known to have microdeletions of chromosome 22q11, although this was not assessed in all patients.

Operative Treatment

Patients were divided into 3 groups on the basis of operative strategy (Figure 1). Group 1 patients (n=56, 66%) underwent complete unifocalization of pulmonary blood supply and intracardiac repair in a single stage through a midline approach. Group 2 patients (n=23, 27%) underwent complete unifocalization through the midline, but the VSD was left open. Group 3 patients (n=6, 7%) underwent staged unifocalization through sequential thoracotomies, often during the same hospitalization. The decision to leave the VSD open after complete unifocalization was generally based on clinical assessment of the pulmonary circulation (discussed elsewhere) but in 2 patients was done to avoid prolonged CPB in a patient with chronic lung disease or aortic cross-clamping in a severely cyanotic patient who had been on neosynephrine infusion for 2 weeks. Patients who underwent staged unifocalization had severe distal segmental MAPCA stenoses (n=3), a diffusely scarred mediastinum after previous bilateral thoracotomies (n=1), or significant comorbid conditions, such as biliary atresia (n=1) or severe cyanotic spells with multiple preoperative cardiac arrests (n=1). Of the patients who had undergone prior palliation, 6 were in group 1, 7 were in group 2, and 1 was in group 3.

Unifocalization of Pulmonary Blood Supply

We have described our approach to 1-stage unifocalization of pulmonary blood supply in a previous report.[21] Although the surgical procedure varies from patient to patient because of the heterogeneity of pulmonary blood supply, our approach is based on the importance of controlling all MAPCAs before commencing CPB and performing native tissue-to-tissue reconstruction whenever possible. A number of techniques helped us achieve these goals, as described elsewhere.

The mediastinum was entered through an extended median sternotomy, which was retracted widely to improve exposure. Other investigators have used a bilateral transsternal thoracotomy.[22] MAPCAs were identified and dissected by use of a variety of techniques. The pleural
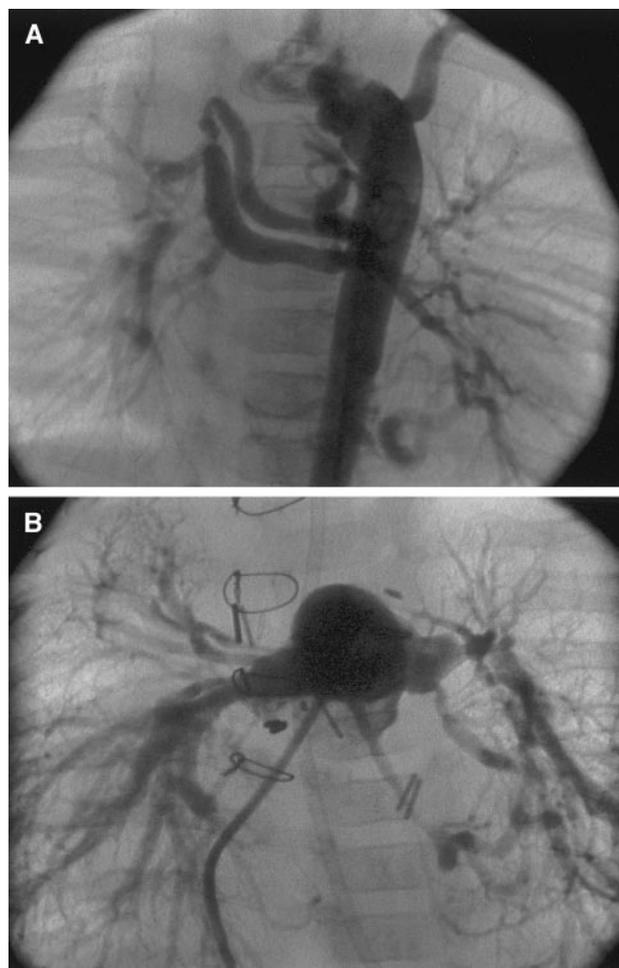
Figure 2. Aortic arch angiogram before (A) and main pulmonary arteriogram after (B) 1-stage complete unifocalization in 2-month-old infant with 5 MAPCAs and 1.5-mm confluent true PAs. VSD was left open in patient after mean PA pressure during intraoperative pulmonary flow study was measured at 30 mm Hg. Postoperative pulmonary angiogram demonstrates complete perfusion of both lungs.

spaces were opened widely anterior to the phrenic nerves, and the lungs were lifted out of their pleural cavities, facilitating identification of collaterals at their origins. MAPCAs from the upper descending aorta were identified and mobilized in the subcarinal space by dissecting between the right superior vena cava and the ascending aorta through the floor of the pericardial reflection in the transverse sinus and the posterior mediastinal soft tissues. MAPCAs arising from the aortic arch, brachiocephalic vessels, or coronary arteries were also exposed and dissected. Immediately before CPB was initiated, all MAPCAs were ligated at their origin to achieve controlled perfusion.

As many MAPCAs as possible were ligated, mobilized, and unifocalized without CPB. With the ligation of each MAPCA, the decrease in arterial oxygen saturation was assessed by pulse oximetry, so it was possible to unifocalize MAPCAs without bypass until desaturation approached a compromising level. At this point, partial CPB was instituted at moderate hypothermia with the heart beating, and the unifocalization was completed. A calcium-supplemented blood prime was used to maintain normal cardiac function. Unifocalization was performed with an emphasis on native tissue-to-tissue anastomosis, which required being aggressive in mobilizing MAPCAs with maximum length, creative in rerouting MAPCAs, and flexible in reconstruction of the neo-PA system. Avenues for collateral rerouting were developed as necessary by opening the pleurae posterior to the phrenic

Demographic, Diagnostic, and Outcome Data in the 3 Clinical Groups

	Total	Group 1	Group 2	Group 3
Patients, n	85	56	23	6
Age				
Median	7 mo	6.5 mo	7 mo	8.7 mo
Range	10 d–37 y	10 d–37 y	1.5 mo–12 y	2 mo–2 y
Infants, n	57	36	16	5
Previous palliative operations,* n	14	6	7	1
MAPCAs, n				
Median	4	4	4	4
Range	1–7	1–7	1–6	3–6
Absent true PAs,* n	14	8	3	3
Early deaths, n	9	5	4	0
Late deaths, n	7	7	0	0

*Number of patients.

nerves in the hilar regions and the subcarinal space through the transverse sinus. A variety of peripheral and central reconstructive techniques were used, including side-to-side or oblique end-to-side anastomosis of MAPCAs to other MAPCAs or to peripheral true PAs, anastomosis of true PAs to an aortic button giving off multiple unobstructed MAPCAs, extended onlay or side-to-side anastomosis of MAPCAs to the central PAs, end-to-end or end-to-side anastomosis of MAPCAs to a central conduit, allograft patch augmentation of distal MAPCA stenoses or of the reconstructed central PAs, and in rare cases reconstruction of central PAs with an allograft conduit. These techniques were used as necessary in a given patient and frequently combined, depending on the anatomy. Direct tissue-to-tissue anastomoses were achieved by bringing MAPCAs through the transverse sinus, or below or above the hilum, with as much of the MAPCA length used as possible. All MAPCAs were incorporated into the reconstruction with these methods, including those that provided dual supply to a lung segment along with a true PA, to augment the neo-PAs with as much native tissue as possible (Figure 2).

In 70 of 79 group 1 and 2 patients, blood flow to the unifocalized PAs was provided via a valved allograft conduit from the RV to the central PAs. In 5 patients (4 in group 2), a transannular outflow tract patch was performed instead, and in 4 others (all group 2), a central systemic-to-PA shunt was used to supply the unifocalized PAs to avoid aortic cross-clamping. After unifocalization, the distal end of the conduit was anastomosed to the central PAs. If necessary, allograft tissue from the conduit was extended distally to augment the central PAs. Before completion of the proximal anastomosis, intracardiac repair was performed.

Intracardiac Repair
The issue of whether to close the VSD at the time of unifocalization is critical to successful repair. When 1-stage unifocalization is performed, it may be difficult to assess whether the VSD should be closed or left open, as we have discussed in a separate report.[23] We have found the most useful approach to be an intraoperative pulmonary flow study that we developed to estimate the resistance of the unifocalized PA bed.[23] After completion of the unifocalization, a pressure-monitoring catheter and perfusion cannula were placed into the distally attached valved conduit, and the cannula was snared. While the left atrium was vented vigorously, the lungs were perfused from the pump with gradually increasing flow equivalent to ≥ 1 estimated cardiac index (2.5 L · min^{-1} · m^{-2}). If the mean PA pressure was <30 mm Hg, the VSD was closed. Otherwise, it was typically left open.

After completion of the unifocalization and flow study in patients who underwent RV outflow tract reconstruction with a valved conduit, the aorta was cross-clamped, and cardioplegia was administered. A longitudinal infundibulotomy was performed, and the

hypertrophied muscle bundles were resected. In group 1 patients, the VSD was closed with an autologous pericardial patch. If an interatrial communication was present, it was closed partially through a right atriotomy to leave a small unidirectional communication for decompression of the right side of the heart. In some cases with intact atrial septum, a small 1-way interatrial communication was created. RV outflow tract reconstruction was completed by anastomosing the conduit to the infundibulotomy.

Statistical Analysis
Perioperative and follow-up data were collected retrospectively by review of hospital records and contact with the referring cardiologist. Cross-sectional follow-up was performed in early 1998. Data are presented as median and range unless otherwise specified. Statistical analysis was performed to assess for demographic differences between patients in the different groups and for factors correlating with early mortality, poor survival over time, and distribution of pulmonary blood flow on follow-up perfusion scan. Independent variables analyzed included age, prior operations, number of MAPCAs, absence of true PAs, chromosome 22q11 deletion/DiGeorge syndrome, treatment group, CPB time, aortic cross-clamp time, postoperative RV-to-LV pressure ratio (group 1), and early reoperation. Dichotomous variables were analyzed by use of the χ^2 test. Comparison of mean values of continuous variables between 2 groups was performed by independent-samples t test. Actuarial survival analysis was performed by the Kaplan-Meier product-limit method. Cox proportional-hazards regression was used for analysis of factors correlating with poorer survival over time.

Results
Patients in group 2 or 3 (combined) were significantly more likely to have undergone previous interventions than patients in group 1 ($P=0.05$). Age, number of MAPCAs, and frequency of absent true PAs did not differ between groups.

Early Results
The intraoperative pulmonary flow study was used in 37 patients, 30 of whom had the VSD closed on the basis of the data obtained. In the 30 patients who had the VSD closed, the mean PA pressure after bypass ranged from 15 to 30 mm Hg (median, 21 mm Hg). In 28 of these 30 patients, the mean PA pressure measured after closure of the VSD and separation from bypass was within 20% and 5 mm Hg of the PA pressure measured during the flow study. In the other 2 patients, in whom pressures during the flow study were 15

and 17 mm Hg, postbypass mean PA pressures were 21 and 25 mm Hg, respectively. In group 1 patients, the ratio of RV to left ventricular (LV) pressure in the operating room ranged from 0.25 to 0.8 (median, 0.44) in all but 1 patient, who underwent fenestration of the VSD patch during the same operation after the ratio was measured at 1.2. The duration of cardiopulmonary bypass (CPB) ranged from 142 to 489 minutes (median, 265 minutes) in group 1 patients and from 126 to 350 minutes (median, 266 minutes) in group 2 patients. Cardioplegic arrest in group 1 patients ranged from 19 to 165 minutes (median, 58 minutes).

Outcomes by treatment group are summarized in the Table and Figure 1. There were 9 perioperative deaths (10.6%), including 6 infants (10.5% of infants). Of the deaths, 5 occurred in group 1 patients and the remaining 4 in group 2 patients. Causes of death were multiorgan failure in 5 patients, sepsis in 1, coagulopathy in 1, pacemaker failure in 1, and ventricular arrhythmia in 1. The only independent variable to correlate significantly with perioperative mortality was duration of CPB, which was longer in patients who died than in survivors (389 ± 82 versus 256 ± 70 minutes, $P=0.003$). Reoperation in the early postoperative period was performed in 18 patients. Four patients required reoperation for an incorrect decision about VSD closure: 3 patients in group 2 underwent VSD closure within 16 days of unifocalization, and 1 patient in group 1 required takedown of the VSD patch on the first postoperative day. Since adoption of the intraoperative PA flow study, no patients have required early reoperation to close/open the VSD. Exploration for postoperative bleeding was performed in 7 patients. Five patients underwent unilateral diaphragmatic plication. Two patients required aortopexy for postoperative tracheal compression. Two patients required reoperation for persistent chylothorax. Other early complications in survivors included transient liver failure in 4 patients, cardiac tamponade in 2, sepsis in 1, *Pseudomonas* pneumonia in 1, intraventricular hemorrhage in 1, and unilateral vocal cord paralysis in 1. There were no other clinically evident neurological complications.

Median duration of ventilatory support was 5 days (range, 1 to 33 days). One patient required tracheostomy after failure to wean from mechanical ventilation as a result of bronchial compression and persistent bronchomalacia. Postoperatively, patients remained in the intensive care unit for a median of 6 days (range, 2 to 90 days) and in hospital for a median of 15 days (range, 5 to 100 days).

Follow-Up

Cross-sectional follow-up ranged from 1 to 69 months (median, 22 months). During the follow-up period, there were 7 late deaths, all in group 1 patients, 6 of whom were <6 months of age at the time of repair. Death was due to sepsis in 4 patients and unknown causes in 2 (referred from foreign countries) and occurred after reoperation for conduit replacement in 1. Actuarial survival was 84% at 1 year and 74% at 4 years (Figure 3A). Factors that correlated with poorer survival over time included longer duration of CPB (groups 1 and 2, $P<0.001$), higher RV-to-LV pressure ratio in the early postoperative period (group 1 only, $P=0.005$), and chromo-
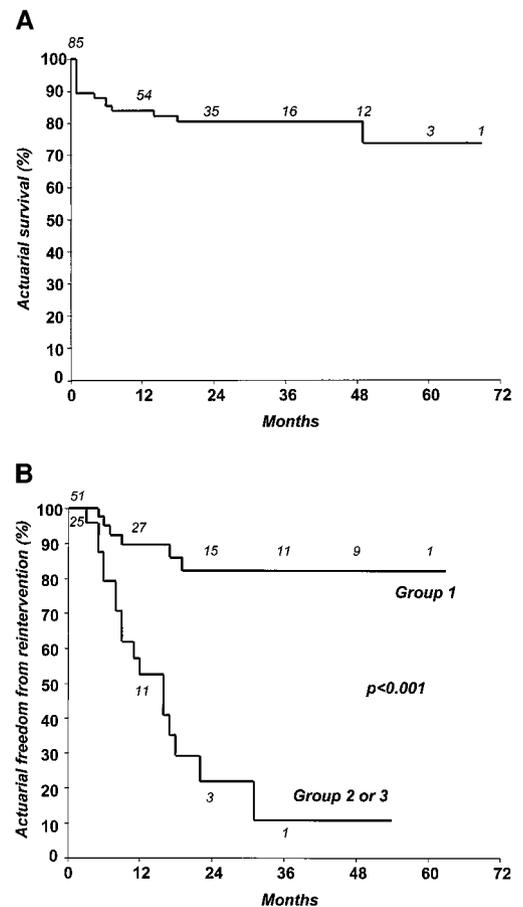
Figure 3. A, Actuarial survival after initial surgical procedure at our institution in 85 patients with pulmonary atresia, VSD, and MAPCAs managed between July 1992 and April 1998. B, Actuarial freedom from reintervention (catheter or surgical) on neo-PAs (does not include isolated conduit changes or pulmonary aneurysmorrhaphy) among early survivors (n=76) after initial unifocalization procedure at our institution.-

some 22q11 deletion ($P=0.05$). None of the other factors analyzed were significant predictors of poor survival.

Of the 19 surviving group 2 patients, 13 underwent completion of the repair, with VSD closure a median of 8 months (range, 6 days to 24 months) after unifocalization. All 6 patients in group 3 underwent complete unifocalization of both lungs, and 2 received complete repair, with VSD closure 8 and 11 months, respectively, after the initial unifocalization procedure. At the time of cross-sectional follow-up, 10 patients in groups 2 and 3 were awaiting closure of the VSD at a median of 15 months after the initial unifocalization procedure. Among early survivors, actuarial survival with complete repair was 83% at 1 year and 88% at 2 years.

Three patients had the RV-PA conduit replaced between 18 and 20 months after unifocalization, 1 at the time of VSD closure in a group 2 patient. Two other patients had resection of a pseudoaneurysm of the RV outflow tract and placement of an RV-PA conduit 3 and 9 months, respectively, after initial outflow tract reconstruction with a transannular patch. Of early survivors, 24 (32%) underwent a total of 36 neo-PA reinterventions. Of these patients, 6 were in group 1, 14 in group 2, and 4 in group 3. Of the 24 patients, 11 were <1 year
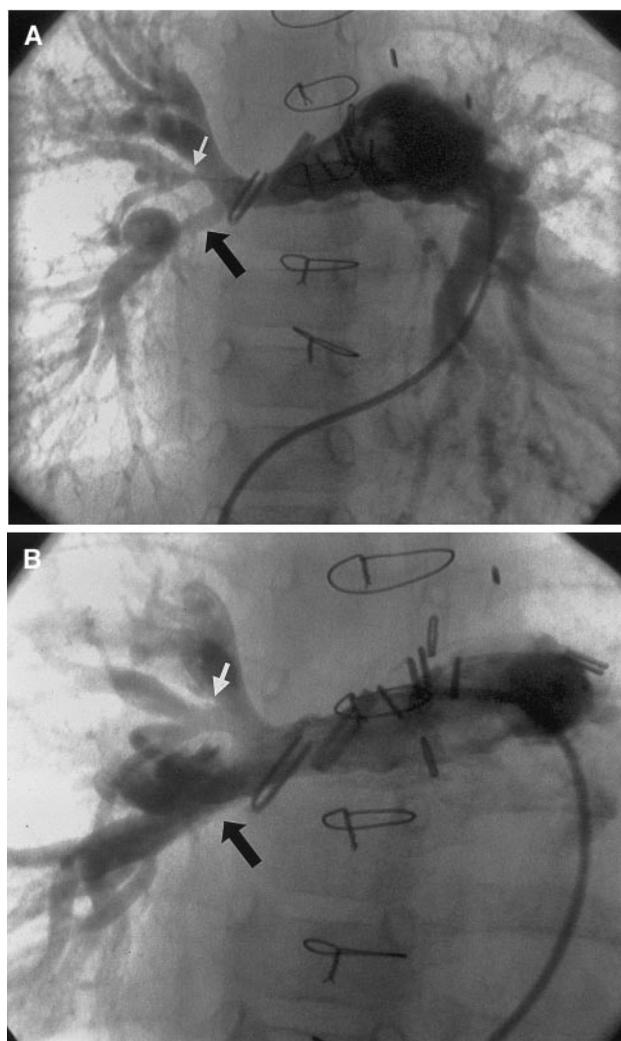
Figure 4. Pulmonary arteriograms before (A) and after (B) balloon dilatation of stenoses in unifocalized right PA (arrows) 2 years after surgery in patient who underwent complete unifocalization without VSD closure at 1.5 months of age. Stenoses in right middle and lower lobe branches were both dilated successfully.

of age at repair. Of the 11 infants who underwent reintervention, 4 were in group 1, 6 in group 2, and 1 in group 3. Eight patients, including 6 in groups 2 and 3, had multiple reinterventions. Transcatheter dilatation procedures were performed in 22 patients, and surgical pulmonary arterioplasty was done in 11 (Figure 4). The median duration from unifocalization to reintervention on the PAs was 8 months (range, 3 to 55 months). In 6 group 2 patients, reintervention on the neo-PAs was performed at the time of VSD closure and/or the catheterization performed immediately before reoperation for VSD closure. Actuarial freedom from neo-PA reintervention among early survivors was 75% at 1 year and 42% at 5 years (Figure 3B) and was significantly higher over time in group 1 than group 2 or 3 patients ($P<0.001$).

Among the 76 early survivors, 60 had ^{99m}Tc lung perfusion scans performed after complete unifocalization. Median percent of pulmonary blood flow to the right lung was 58% (range, 11 to 88%) and to the left lung was 42% (12% to

89%). None of the factors analyzed were found to correlate with distribution of pulmonary blood flow.

Discussion

In 1995, we reported our initial experience with 1-stage unifocalization of MAPCAs through a midline approach in 11 patients.[21] We have subsequently performed unifocalization procedures in 74 additional patients with pulmonary atresia, VSD, and MAPCAs. Since our initial report, in which all 11 patients had complete unifocalization and repair performed in a single stage, our strategy has matured, and we currently use 3 primary approaches. Complete 1-stage unifocalization is still performed in almost all patients (93%), and most (66%) undergo complete repair in a single stage. However, in 29% of patients who were unifocalized in a single stage, VSD closure was postponed to a second operation. The third approach, in which sequential unilateral unifocalization procedures are performed, followed later by intracardiac repair, is reserved for patients with multiple significant distal MAPCA stenoses or with significant comorbid factors that contraindicate CPB.

Outcomes with these strategies have been good, with 10.6% early mortality and 80% actuarial survival at 3 years in an unselected cohort of patients. Among early survivors, actuarial survival with complete repair was 88% at 2 years. Although all 7 late deaths were in group 1 patients, RV hypertension was not a contributing factor in any of these deaths. Early and intermediate outcomes have been almost identical among infants, who constitute 67% of our patients. Although early and intermediate survival has been favorable, reinterventions on the neo-PAs are frequently necessary. During follow-up, 24 patients underwent 36 reinterventions on the neo-PAs, with 8 patients undergoing multiple reinterventions. Actuarial reintervention-free survival was 42% at 5 years.

The integrity of the pulmonary vascular bed is a major issue in the management of pulmonary atresia with MAPCAs. Most staged approaches use circumferential nonviable conduits in the central and peripheral pulmonary circulation, which limits growth potential and the ability to perform complete repair in early infancy. Our approach to 1-stage unifocalization depends on aggressive and creative reconstructive techniques to maximize native tissue-to-tissue connections and avoid nonviable conduits. Thus, it is important to emphasize some of the technical measures described in the Methods section. Most MAPCAs originate from the descending thoracic aorta, often in the subcarinal space. An important approach for accessing and rerouting such MAPCAs to facilitate tissue-to-tissue anastomoses is to dissect between the right superior vena cava and the ascending aorta, then through the floor of the pericardial reflection in the transverse sinus and the posterior mediastinal soft tissues. In addition, the pleurae are opened widely both anterior and posterior to the phrenic nerves, allowing flexibility in perihilar rerouting of MAPCAs. Although it is occasionally necessary to augment the neo-PAs peripherally with allograft patches, these techniques have helped us avoid circumferential conduits in the periphery in all patients.

There has been limited experience with unifocalization techniques based on the principle of maximizing native tissue-to-tissue anastomoses. Thus, it is not known how reconstructed neo-PAs, consisting of unifocalized native PAs and MAPCAs, will develop, given the variable morphology of the vessels before unifocalization and the tendency for MAPCAs to become stenotic in the nonunifocalized state. The fact that one third of survivors have required some form of neo-PA reintervention concerns us somewhat. However, 18 of the 24 patients who underwent reintervention were in groups 2 and 3. These patients did not all necessarily require reintervention but sometimes had procedures performed to correct minor stenoses at the time of VSD closure, a second unifocalization, or catheterization before such reoperations.

Another important aspect of our approach is the issue of when to close the VSD. Since initiating the 1-stage approach, we have improved our ability to determine whether VSD closure should be performed at the time of unifocalization, an improvement that has had clear clinical consequences. Since early 1996, 37 patients have undergone the intraoperative pulmonary flow study that we initially described in a previous report.[23] In addition to these patients, the decision was made to leave the VSD open in certain group 2 patients without the flow study, either because of extenuating clinical circumstances (eg, chronic lung disease and the desire to minimize bypass time) or when the unifocalized neo-PA bed was very diminutive. Since we adopted the intraoperative flow study, no patients have required early reoperation for VSD closure/reopening, whereas it was necessary to close or reopen the VSD in the early postoperative period in 4 patients before the flow study was initiated. Among the 30 group 1 patients who underwent VSD closure after an intraoperative flow study predicted adequate pulmonary vascular resistance, postoperative PA pressures were similar to those obtained during the flow study in all patients, and there was only 1 perioperative death. Follow-up catheterization has not been indicated in most of these patients, so the accuracy of the intraoperative flow study for estimating long-term pulmonary hemodynamic after VSD closure cannot yet be assessed. However, the fact that none of these 30 patients have evidence of pulmonary hypertension clinically or echocardiographically is encouraging.

It is difficult to compare our results to those of previous series in a meaningful fashion, primarily because of the incomparability of denominators. Our series is unique insofar as a substantial proportion of our patients were infants, which probably affects the aptness of comparison with older series in that the highest rate of attrition (with or without surgery) in patients with this lesion is in infancy.[20] Thus, patients who survive beyond infancy are a select population. This is inevitably true with congenital heart disease, and our own cohort is selected as well; we have had 4 patients (3 infants) scheduled for repair who died before surgery. However, our group of patients is likely more representative of the overall population of individuals born with this anomaly than are series of older patients. As discussed elsewhere, our approach is based on the belief that repairing this lesion early in infancy provides the best chance for survival with a normalized circulation. Nonetheless, there is likely a subset of patients who will not survive beyond infancy even with surgery, and they may appear in our series as early or late mortality.

In conclusion, our approach to the management of pulmonary atresia with VSD and MAPCAs is applicable in >90% of patients, even those with absent true PAs, and yields good functional results. Two thirds of patients undergo complete repair during the same operation. There nevertheless remains room for improvement; actuarial survival at 4 years is 74%, and there has been a significant rate of neo-PA reintervention. These results must be appreciated within the natural history of this lesion, according to which an estimated 65% of patients survive to 1 year of age and 50% to 2 years, regardless of surgical intervention.[20] As our experience with this approach increases, we are improving our understanding of how to manage these patients better and anticipate that outcomes will continue to improve. Several centers have recently reported their initial experience with 1-stage unifocalization,[22,24] which will stimulate further progress in the treatment of this difficult lesion. Results with staged repair are also improving, which may bode well for an integrated approach in certain complex patients.

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