CONGENITAL HEART

Surgical Repair of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collaterals With Absent Intrapericardial Pulmonary Arteries

Sergio A. Carrillo, MD, Richard D. Mainwaring, MD, William L. Patrick, BS, Holly D. Bauser-Heaton, MD, PhD, Lynn Peng, MD, V. Mohan Reddy, MD, and Frank L. Hanley, MD

Divisions of Pediatric Cardiac Surgery and Pediatric Cardiology, Lucile Packard Children's Hospital, Stanford University, Stanford, California

Background. **One anatomic variant of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) is characterized by the absence of intrapericardial pulmonary arteries. This anatomy obviates the possibility of incorporating the pulmonary arteries for reconstruction or palliative procedures. The purpose of this study was to evaluate the surgical results in patients undergoing repair of PA/VSD/MAPCAs with absent pulmonary arteries.**

Methods. **This was a retrospective review of 35 patients who underwent surgical repair of PA/VSD/MAPCAs with absent pulmonary arteries between 2007 and 2014. The median age at the time of surgery was 3.4 months, and the median weight was 4.9 kg. All patients underwent unifocalization of MAPCAs, with an average of 3.5 ± 1.4 MAPCAs per patient.**

Results. **Twenty-eight of the 35 patients (80%) underwent complete single-stage surgical repair, including unifocalization of MAPCAs, VSD closure, and right ventricle to pulmonary artery conduit. After complete repair, the average right ventricular to aortic pressure ratio was 0.33 ± 0.07. There were no deaths in this subgroup. Seven patients (20%) were not deemed suitable candidates for VSD closure after their unifocalization procedure, and therefore underwent palliation with a central shunt. There was 1 operative death and 1 interim death. Three patients have subsequently undergone complete repair, and 2 are awaiting further evaluation and treatment.**

Conclusions. **The majority of patients with PA/VSD/MAPCAs and absent pulmonary arteries can undergo complete single-stage repair with satisfactory postoperative hemodynamics. These results suggest that unifocalization of MAPCAs can provide a reasonable pulmonary vascular bed in the absence of intrapericardial pulmonary arteries.**

(Ann Thorac Surg 2015;100:606–14)

Pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) is a complex and diverse form of congenital heart disease [1]. One aspect of this diversity is the variable anatomy of the central branch pulmonary arteries [2–5]. Approximately 80% of patients with PA/VSD/MAPCAs have central branch pulmonary arteries, whereas 20% of patients have a complete absence of intrapericardial pulmonary arteries [6–11]. The developmental factors resulting in the presence or absence of central pulmonary arteries in PA/VSD/MAPCAs are currently not well defined. However, the anatomy of the central branch pulmonary arteries has important implications regarding the surgical options for these patients.

The central branch pulmonary arteries have been utilized in the surgical treatment of PA/VSD/MAPCAs in several ways. The branch pulmonary arteries provide a convenient target for shunting procedures. The use of shunts in the treatment of PA/VSD/MAPCAs is not surprising from a historical standpoint, as shunting procedures were also used extensively in the early treatment of tetralogy of Fallot. Several groups continue to advocate the use of either a central shunt or right ventricle to pulmonary artery conduit as the principal method to achieve "pulmonary artery rehabilitation" [12–16]. The central pulmonary arteries can also be utilized in conjunction with unifocalization of MAPCAs for repair of PA/VSD/MAPCAs [6–11, 17–19]. This strategy provides a versatile approach to the management of PA/VSD/MAPCAs and has yielded excellent hemodynamic results [20]. There is currently

Accepted for publication March 30, 2015.

Presented at the Fifty-first Annual Meeting of The Society of Thoracic Surgeons, San Diego, CA, Jan 24–28, 2015.

Address correspondence to Dr Mainwaring, Stanford University School of Medicine, 300 Pasteur Dr, Falk CVRC, Stanford, CA 94305; e-mail: mainwaring@stanford.edu.

0003-4975/$36.00
http://dx.doi.org/10.1016/j.athoracsur.2015.03.110

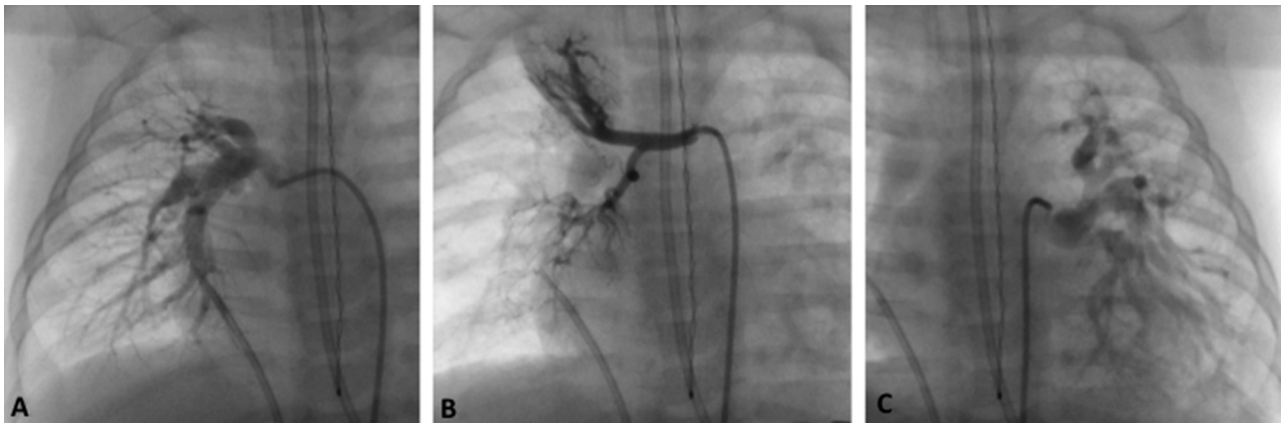

Fig 1. Preoperative angiogram of a typical patient with pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) and absence of intrapericardial central pulmonary arteries. The angiogram demonstrates two MAPCAs to the right lung (A, B) and one large MAPCA supplying the entire left lung (C). There is no filling of a central pulmonary artery on any of the injections. The absence of central branch pulmonary arteries was subsequently confirmed at surgery. This patient was able to undergo a single-stage complete repair.

a spirited debate in the literature regarding the optimal management of patients with PA/VSD/MAPCAs in the presence of central branch pulmonary arteries [21, 22].

For the 1 in 5 patients with PA/VSD/MAPCAs and complete absence of intrapericardial pulmonary arteries, MAPCAs provide the sole source of pulmonary blood flow [8, 9, 19]. This anatomy precludes all of the shunting options because there are no central branch pulmonary arteries to serve as receiving targets for a shunt. In this circumstance, unifocalization of MAPCAs provides the only viable surgical option. There are many published series reporting the results of unifocalization and complete repair, but most have reported a collective experience and have not made a distinction between patients with and patients without intrapericardial pulmonary arteries. The few studies that have analyzed the impact of central pulmonary arteries on outcomes after unifocalization have had mixed results. Carrotti and associates [8] reported that the presence of central pulmonary arteries conferred a benefit to patients undergoing unifocalization and complete repair, whereas Davies and colleagues [10] and our group at Stanford University have reported no significant difference [6, 7].

To date, there has been no study focused solely on patients with PA/VSD/MAPCAs and absent intrapericardial pulmonary arteries. The purpose of the present study was to address this deficiency in the literature by summarizing our experience with repair of PA/VSD/MAPCAs with absent intrapericardial pulmonary arteries.

Material and Methods

This study was approved by the Institutional Review Board at Stanford University. Patients were identified through the cardiac database, and the medical records were subsequently reviewed.

The current study summarizes our experience with 35 patients who were born with PA/VSD/MAPCAs and absent intrapericardial pulmonary arteries. All patients underwent preliminary cardiac catheterization to delineate the anatomy of the MAPCAs (Fig 1). The cardiac catheterization also identified the absence of intrapericardial pulmonary arteries, a finding that would subsequently be confirmed at surgery in all patients. The patients in this study had surgical treatment between 2007 and 2014. There were 20 male and 15 female patients; 14 patients had deletion of chromosome 22q11. The median age at the time of surgery was 3.4 months (range, 1 to 122). The median weight at the time of surgery was 4.9 kg (range, 2.3 to 31.9 kg).

All patients underwent unifocalization of all available MAPCAs using the techniques that have been previously described [6, 7]. A flow study was performed in select patients to evaluate the pulmonary vascular physiology after the unifocalization procedure. Our current criteria for passing the flow study are a mean pulmonary artery pressure less than 25 mm Hg at a flow rate to the lungs of 3 $L \cdot min^{-1} \cdot m^{-2}$ [23]. The patients who were deemed suitable candidates for complete repair underwent VSD closure with an autologous pericardial patch and closure of the atrial septal defect when present. Aortic homografts were utilized to establish continuity of the right ventricle to the reconstructed MAPCAs. Two transthoracic pressure lines were placed at the conclusion of the procedure to monitor left atrial and right ventricular pressures, and representative values were recorded after separation from cardiopulmonary bypass. Patients who were not deemed suitable for complete repair had placement of a central shunt from the ascending aorta to unifocalized MAPCAs.

Statistical results are reported as the mean ± SD. Actuarial analysis was performed comparing patients who underwent complete repair versus patients who

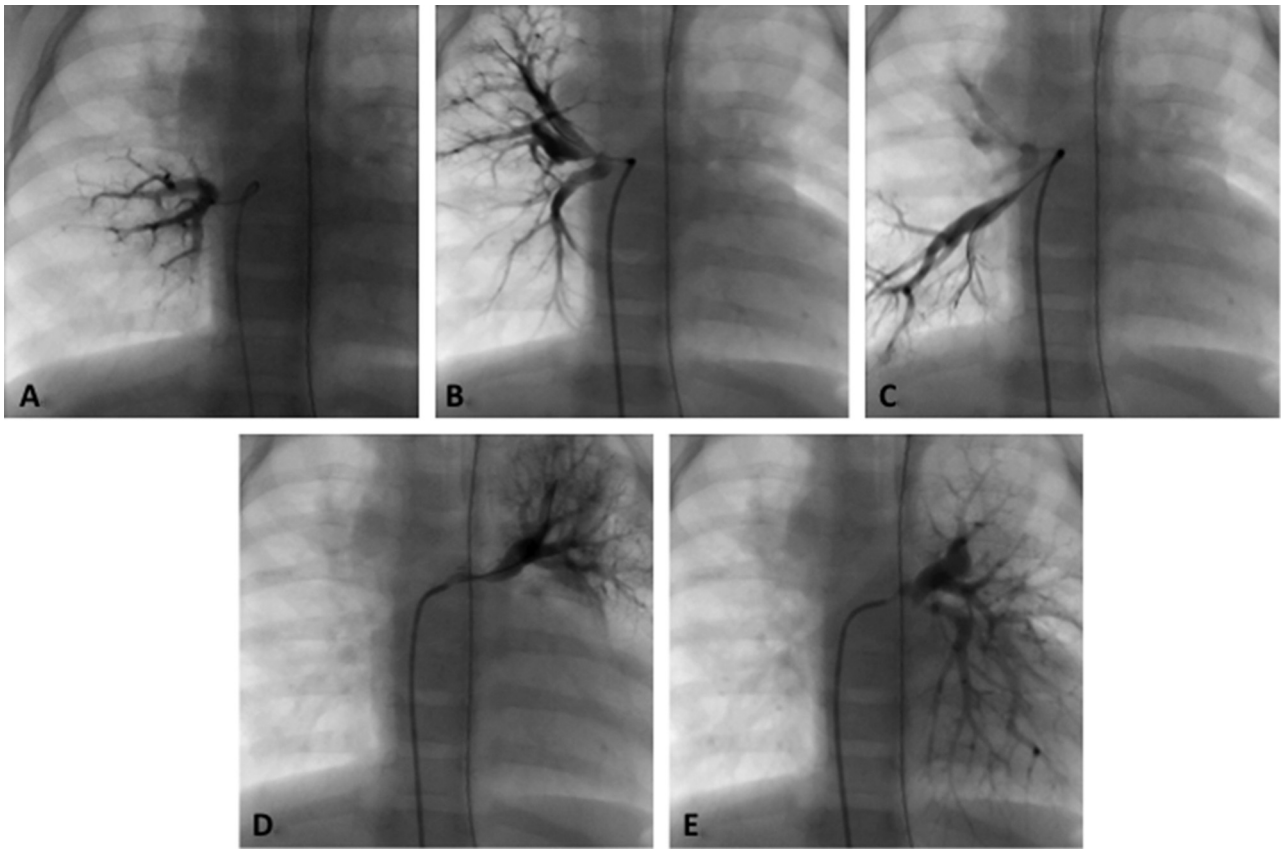

Fig 2. Preoperative cardiac catheterization of a second patient with pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) and absent intrapericardial central pulmonary arteries. The angiograms demonstrate a total of 5 MAPCAs, with three to the right lung (A, B, C) and two to the left lung (D, E). This patient underwent a single-stage complete repair at 8 months of age.

were shunted. Log rank analysis was performed to compare these two subgroups.

Results

All patients underwent a complete unifocalization of their MAPCAs, with an average of 3.5 ± 1.4 MAPCAs per patient. There was an average of 1.9 ± 1.0 MAPCAs to the right lung and 1.6 ± 0.7 MAPCAs to the left lung. Twenty-five patients had favorable preoperative physiology (saturations greater than 85%) indicative of a well-developed pulmonary vascular bed. In this subset of 25 patients, 1 patient had 1 MAPCA, 8 patients had 2 MAPCAs, 6 patients had 3 MAPCAs, 4 patients had 4 MAPCAs, and 6 patients had 5 MAPCAs (for an average of 3.1 ± 1.1 MAPCAs per patient). An example of a patient with 5 MAPCAs is in shown in Figure 2. These 25 patients proceeded directly to complete repair without an intraoperative flow study. Ten patients had less favorable preoperative physiology (saturations less than 80%), indicating that the pulmonary vascular bed was less well developed. In this subset, 1 patient had 2 MAPCAs, 3 patients had 3 MAPCAs, 2 patients had 4 MAPCAs, 2 patients had 5 MAPCAs, and 2 patients had 6 MAPCAs (average 4.2 MAPCAs per patient, $p < 0.05$ compared with subset of 25 patients). These patients had an intraoperative flow study performed. Three of the 10 patients passed the flow study and proceeded to complete repair, whereas 7 patients did not pass the study. An example of a patient with 5 MAPCAs who did not pass the flow study is shown in Figure 3.

The 28 patients who underwent a single-stage complete repair had an average right ventricular peak systolic pressure of 30 ± 6 mm Hg, with an average aortic peak systolic pressure of 91 ± 10 mm Hg (Fig 4). The calculated right ventricle to aortic peak systolic pressure ratio (RV/Ao) was 0.33 ± 0.07. There was no operative mortality in this cohort of 28 patients.

Seven of the 35 patients (20%) with PA/VSD/MAPCAs and absent intrapericardial pulmonary arteries were not deemed suitable candidates for complete repair. These patients underwent placement of a central shunt from the aorta to reconstructed MAPCAs. There was 1 operative death secondary to development of multisystem organ failure. Six patients were discharged from the hospital, and 3 have subsequently undergone complete repair with postoperative hemodynamics (RV/Ao pressure ratio 0.33) similar to those of the 28 patients who underwent a single-stage repair. There was 1 interim death of a patient who had an infectious complication.

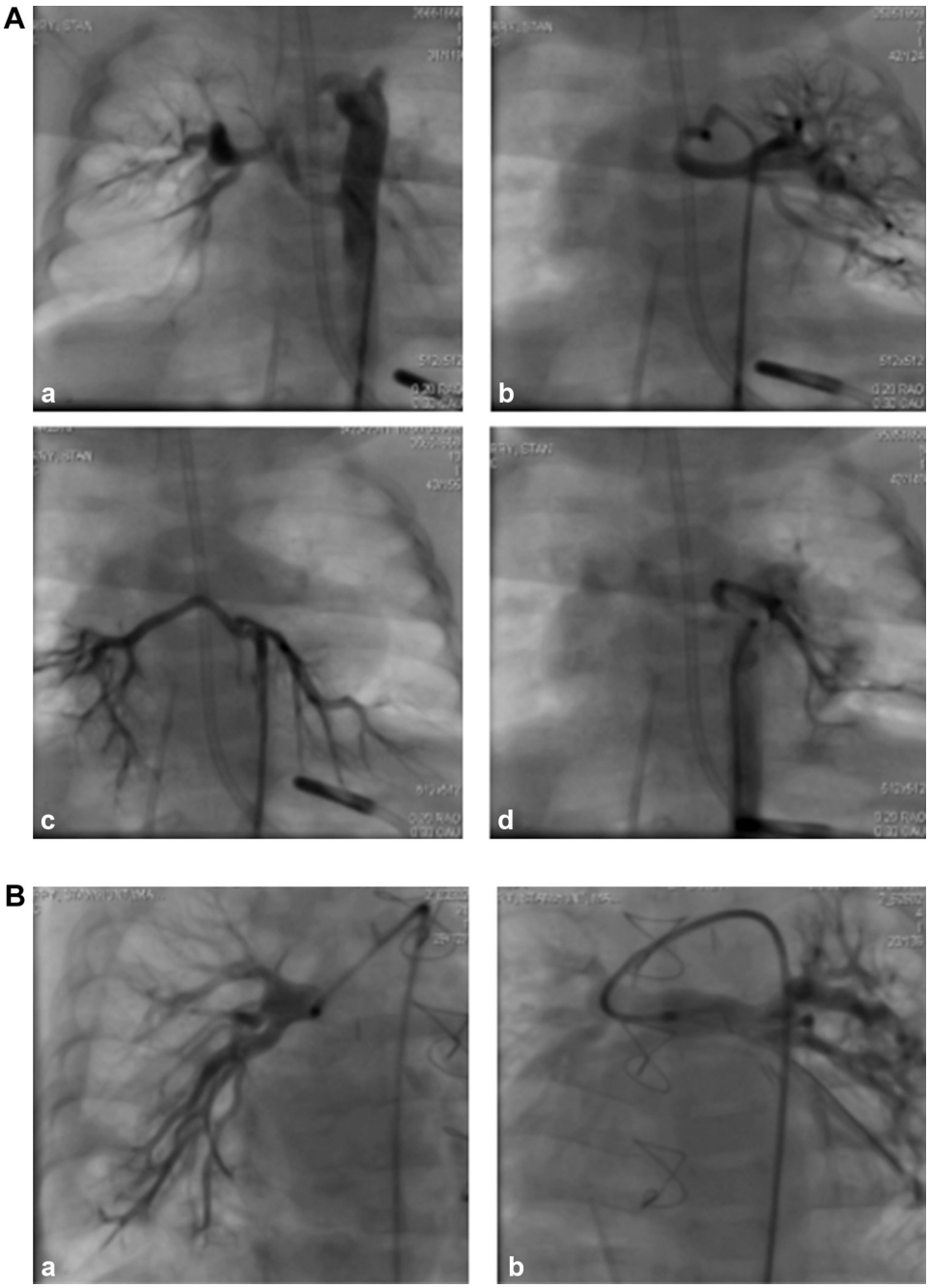

Fig 3. (A) Preoperative angiogram of 1 of the 7 patients who underwent complete unifocalization of their major aortopulmonary collaterals (MAPCAs) but failed the intraoperative flow study and therefore did not undergo complete repair at this operation. The specific patient shown had four MAPCAs (a, b, c, d), with one (c) giving branches to both the right and left sides (so, effectively, three MAPCA branches to the right and two to the left). The angiogram demonstrates the absence of intrapericardial central pulmonary arteries, and that was confirmed at surgery. (B) Angiogram of the same patient after complete unifocalization of the MAPCAs to a central shunt. The two figures demonstrate the separate injections into the right (a) and left (b) lungs. This patient subsequently underwent completion of the repair with closure of the ventricular septal defect and insertion of a right ventricle to reconstructed pulmonary conduit.

Two patients in this subgroup are awaiting further evaluation and treatment. The flow chart for the entire cohort of 35 patients is shown in Figure 5. The actuarial analysis comparing the 28 patients who were able to undergo single-stage complete repair versus the 7 patients who were not able to undergo single-stage complete repair is shown in Figure 6.

The 33 survivors have been followed for an average of 3.7 $\pm$ 2.2 years. To date, 8 patients have undergone subsequent reinterventions on their reconstructed pulmonary vascular beds. Five patients had reinterventions in the cardiac catheterization laboratory. Four of the patients (1 patient with 4 MAPCAs, 2 patients with 5 MAPCAs, and 1 patient with 6 MAPCAs) had postoperative perfusion scans that indicated a shift in pulmonary blood flow distribution. These patients had balloon dilation of relatively discrete areas of proximal stenosis. One patient (originally with 5 MAPCAs) had echocardiographic evidence of increased right ventricular pressure 9 months after single-stage repair. This patient underwent several balloon dilations of peripheral stenoses and a stent in the proximal right pulmonary artery.

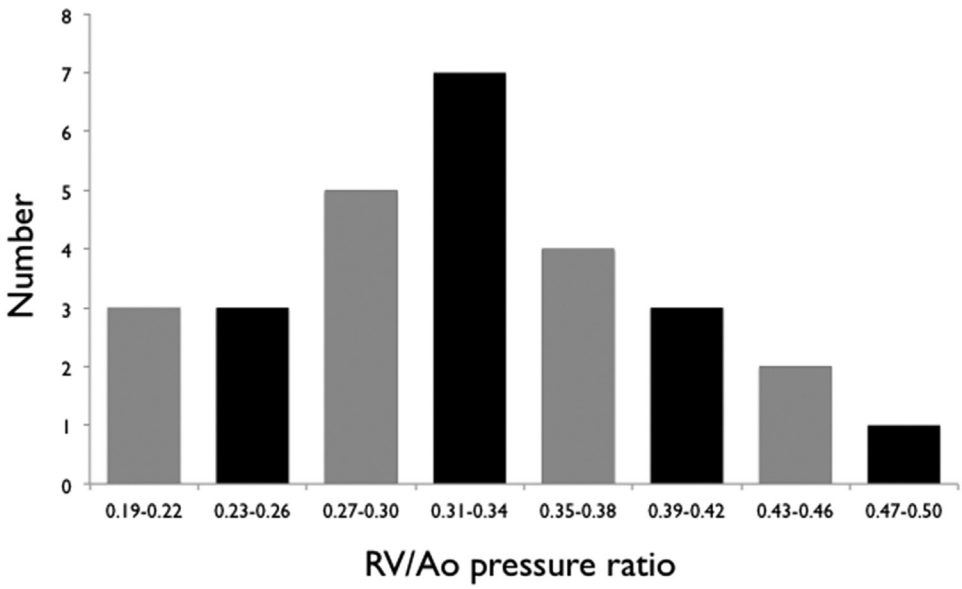

Fig 4. Hemodynamic data for the 28 patients who underwent single-stage complete repair. The average right ventricle (RV) to aortic (Ao) pressure ratio was 0.33 ± 0.07.

Three patients (1 with 4 MAPCAs and 2 with 5 MAPCAs) have undergone reoperation. The first patient had a long segment narrowing of the right lower lobe pulmonary artery. This patient had homograft patch augmentation of this artery, with complete relief of the narrowing and gradient. The second and third patients had proximal pulmonary artery stenosis (one left and one right) in conjunction with conduit dysfunction. These patients had homograft patch augmentation of their proximal pulmonary artery and replacement of the right ventricle to pulmonary artery conduit. The 8 patients who have required reinterventions on their unifocalized pulmonary vascular beds were concentrated at the high end of MAPCAs per patient, with all having either 4, 5, or 6 MAPCAs.

Comment

We have summarized our experience with 35 patients who underwent surgical repair of PA/VSD/MAPCAs in the absence of intrapericardial central pulmonary arteries. These patients had unifocalization of all available MAPCAs, and by definition, the reconstruction was performed without the benefit of any native pulmonary artery tissue. In this study, the percentage of patients who were able to have a single-stage complete repair rate was

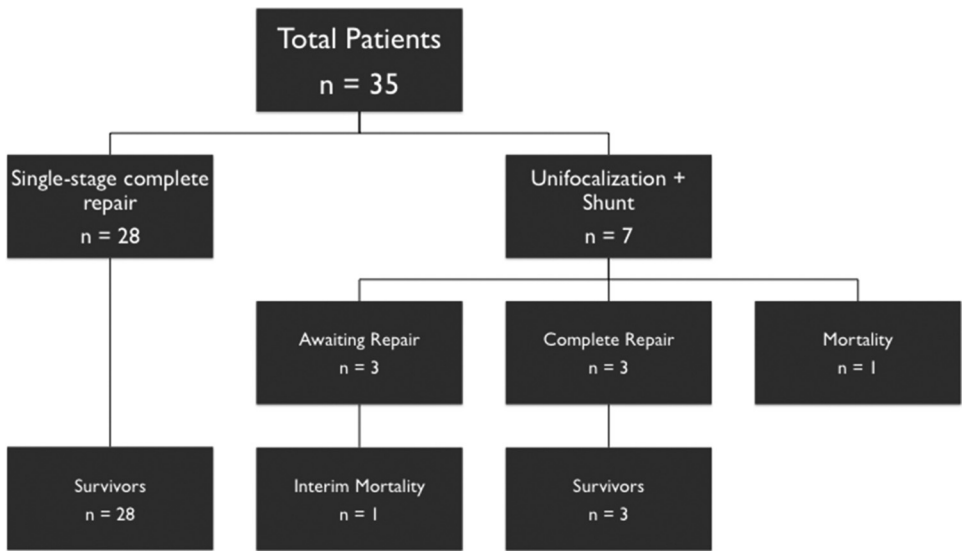

Fig 5. Flow chart demonstrating the results for the 35 patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals and absent intrapericardial central pulmonary arteries.

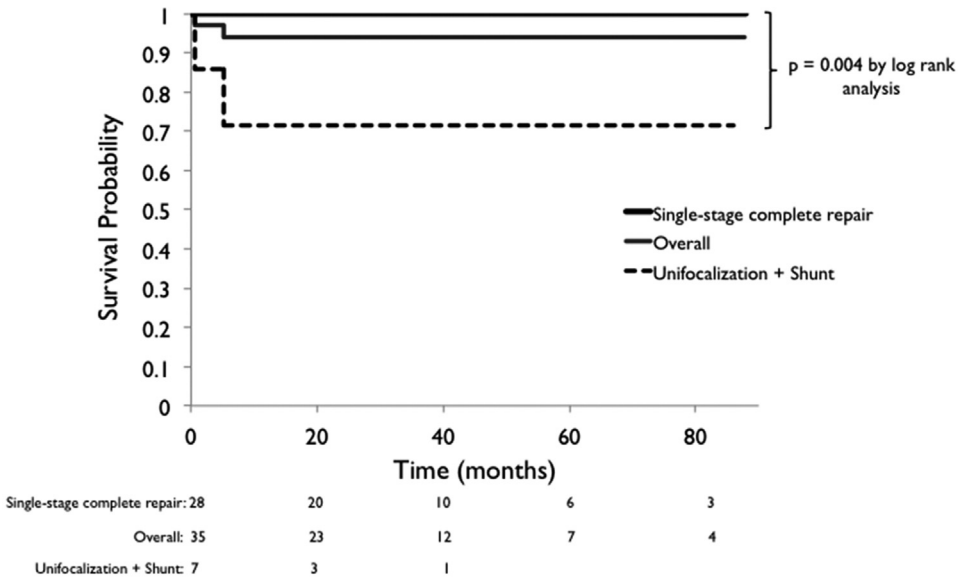

Fig 6. Actuarial analysis comparing the 28 patients who were able to undergo single-stage complete repair (heavy solid line) versus the 7 patients who underwent unifocalization of major aortopulmonary collaterals to a central shunt (dashed line). (Lighter solid line = overall.)

80% (28 of 35), which compares quite favorably with our previously published data for all patients with PA/VSD/MAPCAs [24] in which the percentage of patients who had single-stage complete repair was 56%. These data included both patients with and patients without central intrapericardial pulmonary arteries, and therefore, by inference, the percentage of patients able to achieve single-stage complete repair with central pulmonary arteries would be slightly less than 50%. Therefore, there appears to be a dichotomy between patients with and patients without central intrapericardial pulmonary arteries.

The percentage of patients who are able to undergo a single-stage complete repair is significant, as it provides a proxy regarding the adequacy of the pulmonary vascular bed [25, 26]. Our experience has shown that patients with higher preoperative oxygen saturations (greater than 85%) have a higher likelihood of undergoing single-stage complete repair than patients who do not have these features. The higher oxygen saturations are indicative of increased pulmonary blood flow, and typically correspond to a pulmonary to systemic blood flow ratio of 2:1 or greater at cardiac catheterization. The inference we would draw is that patients who have higher oxygen saturations and larger values of the pulmonary to systemic blood flow ratio have better developed pulmonary vascular beds, and that accounts for the higher likelihood of single-stage complete repair.

The anatomic corollary to increased oxygen saturations is larger sized MAPCAs, which are typically fewer. In this series of patients with PA/VSD/MAPCAs and absent intrapericardial pulmonary arteries, the average number of MAPCAs was just 3.5. The large MAPCAs are relatively straightforward to handle and reconstruct, and also provide ample material for a living tissue to living tissue reconstruction that has ongoing growth potential. The combination of increased oxygen saturations and larger sized MAPCAs was found in 25 of the 35 patients, and for this subset of patients, we no longer perform an intraoperative flow study as we are confident that the patients can have a single-stage complete repair. That is why flow studies were performed in only 10 of the 35 patients in this study.

Thirty-one patients in this series have undergone complete repair (28 single stage and 3 two stage), with an average RV/Ao pressure ratio of 0.33 ± 0.07. This ratio is significantly lower compared with our previously published data, which included nonselected patients with both present and absent central pulmonary arteries. The finding that patients with PA/VSD/MAPCAs and absent central pulmonary arteries have lower RV/Ao pressure ratios than patients with central pulmonary arteries is consistent with the concept that absence of central pulmonary arteries is associated with superior development of the pulmonary vascular bed.

One hypothesis that may account for the more favorable MAPCAs seen in patients with absent native pulmonary arteries relates to the fundamentals of embryologic development. The MAPCAs normally originate from the bronchopulmonary foregut, but involute in response to the subsequent growth and development of the pulmonary vasculature. It is our conjecture that very early atresia of the pulmonic valve results in both a failure of the growth and development of the central pulmonary arteries and failure of the genetic signaling for MAPCA involution. As a consequence, the unaltered MAPCAs remain large and well developed in association with a nicely developed pulmonary vascular bed. Patients with PA/VSD/MAPCAs, central pulmonary arteries, and "dual supply MAPCAs" represent the antithesis of PA/VSD/MAPCAs and absent branch pulmonary arteries. Dual supply MAPCAs

occur where there is normal arborization of the native pulmonary arteries and MAPCAs that supply the same segments of lung. It is our conjecture that this situation is the result of "late" atresia of the pulmonic valve. The consequence is normal arborization of the pulmonary arteries (albeit underdeveloped owing to limited flow) and partial but not complete involution of the MAPCAs. In this setting, not only are the pulmonary arteries hypoplastic, but also the MAPCAs tend to be small and of poor quality. We have previously evaluated our experience with normally arborizing pulmonary arteries and dual supply MAPCAs, which is encountered in approximately 5% of patients, and for whom we advocate an initial aortopulmonary window [27, 28]. In this cohort, the subsequent next-stage complete repair rate was only 12%.

It is evident that the vast majority (70% to 75%) of patients with PA/VSD/MAPCAs fall somewhere between these two extremes. That is most likely the result of atresia of the pulmonic valve somewhere between early and late. The consequence is a wide range of anatomy characterized by varying degrees of pulmonary arborization in combination with some MAPCAs that are dual supply and some that are isolated supply. Our current preference is to perform elective surgical repair for these patients between 3 and 6 months of age. The indications for early repair (which include unremitting heart failure and a ductus or hemitruncus to one lung with MAPCAs to the other) have been outlined in a previous publication [29].

Many groups have now reported their surgical results for repair of PA/VSD/MAPCAs (Table 1). These reports summarize a variety of approaches to and preferences for solving this challenging problem. The percentage of patients undergoing single-stage complete repair has been reported in the literature from 0% to 56%, and ultimately VSD closure in 43% to 85%. Fewer groups have reported their hemodynamic data after complete repair, but for the groups that have, the right ventricle to aortic pressure ratios have ranged from 0.48 to 0.70. Based on the extant literature on tetralogy of Fallot, it is likely that long-term survival will be predicated on the two crucial factors of achieving complete repair with the lowest possible right ventricular pressures.

In summary, the results of this study demonstrate that patients with PA/VSD/MAPCAs and absent central pulmonary arteries had a high incidence of single-stage complete repair and had favorable RV/Ao pressure ratios. These findings are both counterintuitive and contradictory to the widely held belief that the absence of central pulmonary arteries would result in worse outcomes. These results provide further evidence that several subgroups fall under the anatomic diagnosis of PA/VSD/MAPCAs, and these subgroups can be predicted to have different clinical outcomes.

Table 1. Review of Literature on Single-Stage Complete Repair, Ultimate Complete Repair, and Postoperative Hemodynamics of Patients With Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collaterals

First Author	Year Published	Single-Stage Repair	Ultimate Repair	Postoperative RV/LV Ratio
Carrillo	Current study	80%	91%	0.33
d'Udekem	2005	0%	65%	0.62
Ishibashi	2007	0%	81%	0.70
Carotti	2010	48%	77%	0.48
Liava'a	2012	0%	48%	0.64
Griselli	2004	23%	72%	0.60
Song	2009	0%	43%	0.57
Amark	2006	33%	60%	…
Mumtaz	2008	0%	62%	…
Davies	2009	56%	85%	…
De Campli	2010	24%	68%	…
Hibino	2014	0%	76%	…

LV = left ventricle; RV = right ventricle.

References

1. Bull K, Somerville J, Ty E, Speigelhalter D. Presentation and attrition in complex pulmonary atresia. J Am Coll Cardiol 1995;25:491–9.
2. Liao PK, Edwards WD, Julsrud PR, Puga FJ, Danielson GK, Feldt RH. Pulmonary blood supply in patients with pulmonary atresia and ventricular septal defect. J Am Coll Cardiol 1985;6: 1443–350.
3. Rabinovitch M, Herrera-deLeon V, Castenada AR, Reid L. Growth and development of the pulmonary vascular bed in patients with tetralogy of Fallot with or without pulmonary atresia. Circulation 1981;64:1234–49.
4. Jefferson K, Rees S, Somerville J. Systemic arterial supply to lungs in pulmonary atresia and its relation to pulmonary artery development. Br Heart J 1972;34:418–27.
5. Haworth SG, Macartney FJ. Growth and development of pulmonary circulation in pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Br Heart J 1980;44:14–24.
6. 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.
7. Reddy VM, McElhinney DB, Amin Z, et al. Early and intermediate outcomes after repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: experience with 85 patients. Circulation 2000;101:1826–32.
8. Carotti A, Albanese SB, Filleppelli S, et al. Determinants of outcome after surgical treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg 2010;140:1092–103.
9. Griselli M, McGuirk SP, Winslaw DS, et al. The influence of pulmonary artery morphology on the results of operations for major aortopulmonary collateral arteries and complex congenital heart defects. J Thorac Cardiovasc Surg 2004;127: 251–8.
10. Davies B, Mussa S, Davies P, et al. Unifocalization of major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect is essential to achieve excellent outcomes irrespective of native pulmonary artery morphology. J Thorac Cardiovasc Surg 2009;138:1269–75.
11. Amark KM, Karamlou T, O'Connell A, et al. Independent risk factors associated with mortality, reintervention, and achievement of complete repair in children with pulmonary atresia with ventricular septal defect. J Am Coll Cardiol 2006;47:1448–56.

12. Iyer KS, Mee RB. Staged repair of pulmonary atresia with ventricular septal defect and major systemic to pulmonary artery collaterals. Ann Thorac Surg 1989;51:65–72.
13. Duncan BW, Mee RB, Prieto LR, et al. Staged repair of tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg 2003;126:694–702.
14. Mumtaz MA, Rosenthal G, Quereshi A, et al. Melbourne shunt promotes growth of diminutive central pulmonary arteries in patients with pulmonary atresia, ventricular septal defect, and systemic-to-pulmonary collateral arteries. Ann Thorac Surg 2008;85:2079–83.
15. Liava'a M, Brizard CP, Konstantinov IE, et al. Pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals: neonatal pulmonary artery rehabilitation without unifocalization. Ann Thorac Surg 2012;93:185–92.
16. Hibino N, He D, Yuan F, Yu J-H, Jonas R. Growth of diminutive central pulmonary arteries after right ventricle to pulmonary artery homograft implantation. Ann Thorac Surg 2014;97:2129–33.
17. Song SW, Park HK, Park YH, Cho BK. Pulmonary atresia with ventricular septal defects and major aortopulmonary collateral arteries. Circ J 2009;73:516–22.
18. DeCampli WM, Argueta-Morales IR, Zabinsky J, Jannan RL, Burke RP. An institutional approach to, and results for, patient with tetralogy with pulmonary atresia and major systemic-to-pulmonary collateral arteries. Cardiol Young 2010;20(Suppl 3):128–34.
19. Ishibashi N, Shin'oka T, Ishiyama M, Sakamoto T, Kurosawa H. Clinical results of staged repair with complete unifocalization for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Eur J Cardiothorac Surg 2007;32:202–8.
20. Mainwaring RD, Reddy VM, Peng L, Kuan C, Palmon M, Hanley FL. Hemodynamic assessment after complete repair of pulmonary atresia/major aortopulmonary collaterals. Ann Thorac Surg 2013;95:1397–402.
21. Norgaard MA, Alphonso N, Cochrane AD, Menahem S, Brizard CP. d'Udekem Y. Major aorto-pulmonary collateral arteries of patients with pulmonary atresia and ventricular septal defect are dilated bronchial arteries. Eur J Cardiothorac Surg 2006;29:653–8.
22. d'Udekem Y, Alphonso N, Norgaard MA, et al. Pulmonary atresia with ventricular septal defects and major aortopulmonary collateral arteries: unifocalization brings no long-term benefit. J Thorac Cardiovasc Surg 2005;130:1496–502.
23. Mainwaring RD, Sheikh AY, Reddy VM, Punn R, Hanley FL. Surgical results in patients with pulmonary atresia/major aortopulmonary collaterals and Alagille syndrome. Eur J Cardiothorac Surg 2012;42:235–41.
24. Malhotra SP, Hanley FL. Surgical management of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: a protocol-based approach. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009;12:145–51.
25. Grosse-Wortmann L, Yoo S-J, van Arsdell G, et al. Preoperative total pulmonary blood flow predicts right ventricular pressure in patients early after complete repair of tetralogy of Fallot and pulmonary atresia with major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg 2013;146:1185–90.
26. Honjo O, Al-Radi OO, MacDonald C, et al. The functional intraoperative pulmonary blood flow study is a more sensitive predictor than preoperative anatomy for right ventricular pressure and physiologic tolerance of ventricular septal defect closure after complete unifocalization in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals. Circulation 2009;120:546–52.
27. Mainwaring RD, Reddy VM, Perry SB, Peng L, Hanley FL. Late outcomes in patients undergoing aortopulmonary window for pulmonary atresia/stenosis and major aortopulmonary collaterals. Ann Thorac Surg 2012;94:842–8.
28. Mainwaring RD, Reddy VM, Hanley FL. Surgical reconstruction of pulmonary stenosis with ventricular septal defect and major aortopulmonary collaterals. Ann Thorac Surg 2013;95:1417–21.
29. Watanabe N, Mainwaring RD, Reddy VM, Palmon M, Hanley FL. Early complete repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. Ann Thorac Surg 2014;97:909–15.

DISCUSSION

DR JEFFREY P. JACOBS (St. Petersburg, FL): Thank you very much. Clearly, this is a lesion that your institution is known for more than most. While people are formulating some questions, you may have said this, but what is your cutoff to completely septate the heart versus leaving a fenestrated ventricular septal defect (VSD), or not closing the VSD, or reopening the VSD for RV/aortic pressure ratios?

DR CARRILLO: Thank you, Dr Jacobs. As a matter of fact, we do not fenestrate the VSD. What we do in the operating room is do a flow study, and the criteria to pass a flow study is a mean pulmonary arterial pressure of 25 or less at a flow rate to the lungs of 3 L per minute per meter squared.

DR JACOBS: And if it is more than that, you just leave the VSD open?

DR CARRILLO: No. If it is more than that, what we do is create a systemic to pulmonary artery shunt and we leave the VSD open, clearly, yes.

DR JACOBS: So are there any patients where you had the pulmonary artery connected to the right ventricle with the VSD left open?

DR CARRILLO: No. No, none.

DR YVES D'UDEKEM (Melbourne, Australia): Congratulations for this presentation. These results are magnificent. I was particularly impressed by the second case, since you had only 36% of the aortic pressure in the right ventricle, especially since I thought you had lost some of the two inferior major aortopulmonary collaterals (MAPCAs). And I was surprised that you can achieve such low pressure with these distal pulmonary arteries. My feeling is that when you talk about the patients with absent central pulmonary arteries, that encompasses a variation within the same condition, a spectrum of disease, and I think your examples were quite good for that.

You have patients who have a high pulmonary blood flow because they have isolated MAPCAs going to almost normal intrapulmonary branches. These patients can be in heart failure early in life. And obviously these patients should have these intrapulmonary arteries reconnected to the central pulmonary arteries; but you have also patients with very small, multiple collateral arteries. So my question is, did you try to analyze the characteristics of the patients who were failing the single-stage repair, number of MAPCAs, or maybe I was thinking, did you look at the age between the two groups? Because age may be

your surrogate to know whether the patients were in heart failure early after birth, or maybe had multiple small MAPCAs.

DR CARRILLO: Thank you for your comments, and certainly you have done plenty of work on this topic. First of all, we did not delineate. We did not think that the age was a risk factor. The age of these patients were all infants basically. There was just 1 patient who was above infancy, and we do not believe that age is a risk factor for the presence of MAPCA, or the number of MAPCAs.

We did notice that there is a trend toward more difficult reconstruction of patients who have a higher number of MAPCAs; and the reason why is because the more MAPCAs there are, there is less pulmonary vascular bed development. And if the saturations of the patient are appropriate, let us say if their saturations are greater than 85%, we usually proceed with a single-stage complete repair, unless the patient fails an intraoperative flow study. But in those patients, even with the presence of 5 or 6 MAPCAs, with perfect saturations, that is what we would do. If they have lower saturations or they fail a flow study, we will do a shunt.

DR D'UDEKEM: So if I understand, the patients with more MAPCAs are failing more the single-stage repair?

DR CARRILLO: With higher MAPCAs. They tend to fail single-stage repair because they have segmental stenosis.

DR D'UDEKEM: Maybe they are the ones you should not unifocalize, maybe.

DR CARRILLO: Thank you.

DR SITARAM M. EMANI (Boston, MA): Very nice study and impressive results. Maybe I missed the slide. How many patients had catheter-based reintervention on the pulmonary arteries in each series to achieve these results? I think, as you know, when you go from the front to do the inner focalization, you leave a fair amount of arterialized segment attached, and our experience has been that almost universally, these segments tend to stenose over time. So how often, or if any, catheter-based interventions were necessary to achieve these excellent results?

DR CARRILLO: Thank you, Dr Emani, for your comment and your question. We had a total of 8 patients who required reintervention. Five of them underwent endovascular therapies with balloon dilation, and 1 of them had a central right pulmonary arterial stenosis around the placement of a stent. The other 3, on the one surgical reconstruction, there was 1 patient who had a long segment of right pulmonary artery stenosis after the unifocalization, single stage, that underwent augmentation with a patch. And the other 2 who required operation were patients who had conduit dysfunction at the same time with proximal pulmonary artery stenosis. So somewhere, in total, we had 8 reinterventions: 5 dealt with endovascular therapies and 3 surgical.

DR FRANK SCHOLL (Hollywood, FL): Oftentimes, there are significant distances to get some of these together. What is the preferred material if you do not have enough of the native pulmonary artery to bring these together in the middle? What is your technique?

DR CARRILLO: We prefer obviously the use of native tissue, but if that is not possible, what we use is pulmonary homograft tests that would prefer a patch to augment the MAPCAs. But the majority of the time, the MAPCAs can come close to each other, and in that way, you can create a native tissue, posterior anastomosis, and kind of enlarge the anterior portion of it.

DR JACOBS: Thank you very much.