

# Surgical Reconstruction of Pulmonary Stenosis With Ventricular Septal Defect and Major Aortopulmonary Collaterals

Richard D. Mainwaring, MD, Rajesh Punn, 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.** Pulmonary stenosis with ventricular septal defect and major aortopulmonary collaterals (PS/VSD/MAPCAs) is an extremely rare form of congenital heart defect. Although it has been assumed that PS/VSD/MAPCAs would be similar to pulmonary atresia (PA) with VSD/MAPCA, there are currently no data to support this conjecture. This study reviewed our surgical experience with reconstruction of PS/VSD/MAPCA.

**Methods.** This was a retrospective review of 25 patients (14 boys, 11 girls) who were born with PS/VSD/MAPCA and underwent surgical reconstruction. Preoperative pulmonary angiography was used to define the central branch pulmonary arteries and MAPCA. Patients were a median age of 4 months at the first operation.

**Results.** There was one operative death (4%) in this cohort of 25 patients, and complete repair was achieved in the 24 survivors (96%). There were two distinct subgroups of patients: 11 demonstrated cyanosis in the neonatal timeframe and underwent an initial procedure

to augment pulmonary blood flow (+PBF). The remaining 14 patients formed the second group (−PBF). The median age at the first operation was 0.8 months in the +PBF group and 5.2 months in the −PBF group ( $p < 0.005$ ). Complete repair was achieved in 91% of patients in the +PBF group and in 100% in the −PBF group; however, the average number of procedures to achieve complete repair was 2.8 in the +PBF group vs 1.0 in the −PBF group ( $p < 0.005$ ).

**Conclusions.** Outcomes for PS/VSD/MAPCAs as a whole were excellent, with a low surgical mortality and high rate of complete repair. There were two identifiable subgroups with distinctive differences required in their surgical management. These results provide a prognostic outlook for patients with PS/VSD/MAPCAs that can be compared and contrasted with PA/VSD/MAPCAs.

(Ann Thorac Surg 2013;95:1417–21)

© 2013 by The Society of Thoracic Surgeons

Pulmonary stenosis with ventricular septal defect and major aortopulmonary collaterals (PS/VSD/MAPCAs) is an extremely rare form of congenital heart disease [1–3]. PS/VSD/MAPCAs shares many anatomic features found in pulmonary atresia (PA) with VSD and MAPCAs. These overlapping features include (1) a malalignment type VSD, (2) diminutive main and central branch pulmonary arteries, and (3) the presence of MAPCAs which may supply some or all segments of the lung. Because the anatomic features of PS and PA with VSD/MAPCAs are similar, they may also share many physiologic characteristics. This overlap of anatomy and physiology may account for why these two entities heretofore have been virtually indistinguishable from one another.

There is now extant literature on the subject of surgical reconstruction for PA/VSD/MAPCAs [4–9]. It is noteworthy that none of these reports contain a description of the entity of PS/VSD/MAPCAs. It is not likely that patients

with PS/VSD/MAPCAs were never encountered in these previous reports. Therefore, it would appear that the primary focus to this point has been on the development of management algorithms. Because these management strategies applied equally well to PA and PS with VSD/MAPCAs, the distinction between these two entities has not been previously emphasized.

Although PS/VSD/MAPCAs may have similarities to PA/VSD/MAPCAs, there are currently no data in the literature comparing these two entities. The purpose of this study was to review our clinical and surgical experience with reconstruction of PS/VSD/MAPCAs.

## Material and Methods

This study was approved by the Institutional Review Board at Stanford University. Patients were identified through the cardiac database, and their medical records were subsequently reviewed. The time period encompassed was April 2002 to July 2012.

This was a retrospective study summarizing our experience with 25 patients (14 girls, 11 boys) who underwent surgical reconstruction of PS/VSD/MAPCAs. The aver-

Accepted for publication Jan 7, 2013.

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

age at the first operation was  $5.1 \pm 2.4$  months. Five of the 25 children (20%) were diagnosed with DiGeorge syndrome. All of the patients in this study had multiple echocardiograms, and a cardiac catheterization was performed before their operations. The purpose of the cardiac catheterization was to define the pulmonary artery anatomy, origin, and course of the MAPCAs, and coronary artery pattern.

There were two distinct subgroups of patients as defined by their clinical presentation: 11 presented at an early age with clinical cyanosis, as defined by oxygen saturations of 80% or less. The remaining 14 patients had substantially higher oxygen saturations, with an average of approximately 90%.

The 11 patients who presented with early cyanosis all required an initial procedure to augment pulmonary blood flow. From an anatomic standpoint, these patients had “near-atresia” of the pulmonary valve, with an average pulmonary valve z score of  $-10 \pm 1$ . These patients had very limited flow through the pulmonary valve, and derived almost all of their pulmonary blood flow from MAPCAs. As a consequence, echocardiography was able to identify antegrade pulmonary flow across the pulmonary annulus in only 3 patients, whereas cardiac catheterization found antegrade flow in 9. The remaining 2 patients were identified intraoperatively.

An aortopulmonary window was created in 9 of these 11 patients. This is a surgical technique that we have previously described for this patient population [10]. One patient had slightly larger branch pulmonary

arteries, and a Gore-Tex (W.L. Gore and Associates, Flagstaff, AZ) shunt was placed from the aorta to the main pulmonary artery. The final patient in this group underwent patch reconstruction of the right ventricular outflow tract.

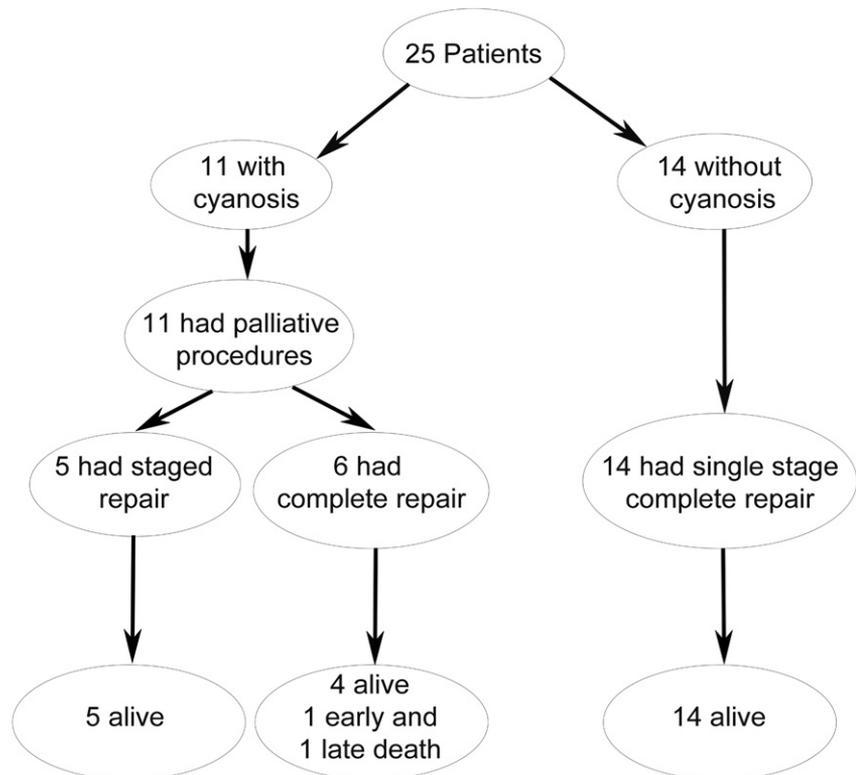
The second subgroup comprised 14 patients who had satisfactory oxygen saturations and therefore were candidates for a single-stage complete repair. These patients had slightly more antegrade flow through the pulmonary valve, and the main and central branch pulmonary arteries were somewhat larger than those in the group with cyanosis. The average pulmonary valve z score was  $-8 \pm 1$ , indicating that almost all of the pulmonary blood flow was still supplied by MAPCAs. Echocardiography identified antegrade flow in 11 patients, and cardiac catheterization visualized antegrade flow in all 14 patients in this subgroup.

Statistical results are reported as the mean  $\pm$  standard deviation. A *p* value of less than 0.05 was deemed statistically significant. Comparison of the two groups was performed using  $\chi^2$  analysis when evaluating rates and a Student *t* test when evaluating absolute numbers.

### Results

There was 1 operative death (4%) and 1 late death (4%) in this cohort of 25 patients with PS/VSD/MAPCAs. Complete repair was achieved in 96% of the patients, which included closure of the VSD and atrial septal defect as well as placement of a conduit from the right ventricle to

Fig 1. Flow diagram shows the 25 patients who underwent surgical treatment for pulmonary stenosis with ventricular septal defect and major aortopulmonary collaterals.



the reconstructed pulmonary arteries. A flow chart for these 25 patients is shown in Figure 1.

In the subgroup of 11 patients who presented with cyanosis, there were no operative deaths for the initial palliative procedure to augment pulmonary blood flow. Six of 11 patients subsequently underwent complete repair as their next surgical procedure, with 1 operative death. Complete repair was ultimately achieved in the remaining 5 patients, but they required between two and four procedures to do so. The average right ventricle-to-left ventricle pressure ratio at complete repair was  $0.42 \pm 0.08$ .

All of the 14 patients who had satisfactory oxygen saturations were able to undergo a single-stage complete repair, with no deaths. The average right ventricle-to-left ventricle pressure ratio was  $0.33 \pm 0.09$  ( $p < 0.05$  vs the subgroup with cyanosis). At the time of complete repair, 12 of these 14 patients underwent unifocalization of one or more collaterals. In addition, 10 of 14 patients had ligation of collaterals that were “dual-supply” and thus did not require unifocalization.

The two groups differed significantly in age at first operation (0.8 vs 5.2 months), average number of procedures to achieve complete repair (2.8 vs 1.0), right ventricle-to-left ventricle pressure ratio (0.42 vs 0.33), long-term survival (83% vs 100%), and incidence of dual-supply MAPCAs (100% vs 28%). The incidence of DiGeorge syndrome was also different in the two subgroups (36% in the cyanotic group vs 7% in the non-cyanotic group).

Coronary artery anomalies were noted in 4 of the 25 patients (16%). Three of these four coronary artery anomalies were the left anterior descending coronary originating from the right coronary. The fourth anomaly was a dual left anterior descending coronary artery, with the duplicate branch originating from the right coronary. Two of the coronary anomalies occurred in the subgroup with cyanosis (representing an incidence of 18%), and two occurred in the subgroup without cyanosis (14%).

The 24 survivors who underwent complete repair have been monitored for an average of  $4.3 \pm 2.0$  years (range, 1 month to 9.5 years). One late death occurred in the group that initially presented with cyanosis. This patient had DiGeorge syndrome and multiple, recurrent infections developed that eventually proved fatal. Four patients have subsequently undergone replacement of their right ventricle-to-pulmonary artery conduit.

## Comment

This report summarizes our experience with 25 patients who underwent surgical reconstruction of PS/VSD/MAPCAs. Complete repair was achieved in 96% of this cohort, with 1 (4%) operative death. Two discernible subgroups of patients were determined by the presence or absence of cyanosis early in life. A comparison of these two subgroups demonstrated significant differences in the age at the first operation, number of procedures required to achieve complete repair, right ventricle-to-left ventricle pressure ratio after complete repair, long-

term survival, and the incidence of DiGeorge syndrome. These results provide a prognostic outlook for patients with PS/VSD/MAPCAs that can be compared and contrasted with PA/VSD/MAPCAs.

The entity of PS/VSD/MAPCAs came squarely to our attention during our research for a report on the surgical creation of aortopulmonary windows [10]. Our indications for aortopulmonary window are quite specific and include clinical cyanosis and diminutive ( $< 2.5$  mm) branch pulmonary arteries that are confluent, have normal arborization, and have dual-supply MAPCAs. We noticed that a disproportionate number of patients in this series had PS/VSD/MAPCAs rather than PA. Although the amount of antegrade flow in these patients is physiologically quite negligible, a link appears to exist between PS/VSD/MAPCAs and the development of dual-supply MAPCAs, an entity that is also not very common. We do not have an explanation to account for this association, but simply point this out as an observation. Once we began to re-review our database, we found 25 patients who anatomically had PS/VSD/MAPCAs, who then became the basis for this report.

Our group has written extensively about the midline surgical approach for unifocalization and complete repair of PA/VSD/MAPCAs [11–13]. This procedure includes providing unobstructed pulmonary blood flow to as many pulmonary segments as possible, patch repair closure of the VSD, and a homograft conduit from the right ventricle to the reconstructed pulmonary arteries. This same approach was used in the patients with PS/VSD/MAPCAs. It should be noted that although the second subgroup of patients did have some measurable amount of antegrade flow through their pulmonary valve, the average z score for this group was  $-8$ , and therefore, these patients were all treated using the same protocol that we use for PA/VSD/MAPCAs, including reconstruction of the right ventricular outflow tract with a valved conduit. It is conceivable that a small subset of patients in this group could be repaired with a transannular patch reconstruction if the pulmonary vascular resistance was sufficiently low to support this arrangement.

We previously reported the clinical outcomes for unifocalization and complete repair in 462 patients at our center [12]. These data demonstrated an overall complete repair rate approaching 90%, with a single-stage complete repair rate of 56%. That report did not make a distinction between patients with PA/VSD/MAPCAs and patients with PS/VSD/MAPCAs, and thus by definition included a mixture of both types of anatomy. Because our institutional series now numbers close to 600 patients, the relative incidence of PA to PS with VSD/MAPCAs appears to be on the order of 25:1. The overall complete repair rate in the present study evaluating only patients with PS/VSD/MAPCAs was 96%. This compares quite favorably with the larger series, particularly because the previous analysis would have included patients with the higher complete repair rate for PS. We thus infer that the diagnosis of PS/VSD/MAPCAs carries a favorable prognosis compared side-by-side with PA/VSD/MAPCAs.

One of the two patient subgroups with PS/VSD/MAPCAs was defined by the presence of cyanosis at an early age. These 11 patients all required an initial palliative surgical procedure to augment pulmonary blood flow. Although 91% of these patients subsequently went on to a successful complete repair, many required multiple procedures to achieve complete repair status. All 11 patients had dual-supply MAPCAs, and yet many subsequently required unifocalization of MAPCAs as a means to address peripheral stenoses in the native pulmonary artery system. We speculate that dual-supply MAPCAs may be a marker for a less well-developed peripheral pulmonary vascular bed due to the competitive flow from the native pulmonary artery system and the MAPCAs. This goes against the conventional wisdom that a dual supply simplifies management because in theory, the MAPCAs can be ligated, occluded, or simply ignored. Our experience would suggest that patients who present with cyanosis could be expected to have a more circuitous pathway to achieve complete repair.

Oxygen saturations were satisfactory in the second subgroup, consisting of 14 patients, and they were therefore candidates for a single-stage complete repair, which was successful in all 14 patients (100%). These results are in marked contrast to those described for the subgroup with cyanosis, who required an initial palliative procedure to increase pulmonary blood flow and had a complete repair rate of only 56% at the subsequent operation. These data suggest that the presence or absence of cyanosis at an early age is an important prognostic indicator for predicting the suitability for a single-stage complete repair.

The incidence of DiGeorge syndrome differed significantly between the cyanotic (36%) and noncyanotic (7%) subgroups. Why there was this disparity and what this difference signifies is unclear. The incidence of DiGeorge syndrome is generally about 30% in most clinical series of patients undergoing unifocalization and complete repair [7]. Some series have reported DiGeorge syndrome is a poor prognostic indicator for late outcomes [5]. To date, however, there are no data to suggest that the genetic defect of 22q11 is a determinant of more or less favorable pulmonary vascular physiology. Future studies may be required to identify whether there is a specific link between the genetic coding and formation of the pulmonary vascular architecture.

In 1996, Kreutzer and colleagues [14] reported the results of a primarily catheter-based approach in 8 patients with PS/VSD/MAPCAs. This strategy began with dilation of the pulmonary valve, which had a z-score of  $-4.0$  before dilation and increased to  $-3.3$  after dilation. Most patients then underwent branch pulmonary artery balloon dilation and coil embolization of aortopulmonary collaterals. All 8 patients were subsequently able to undergo complete surgical repair, with postoperative right ventricle-to-left ventricle pressure ratios averaging about half-systemic. Five of the 8 patients were ultimately able to undergo transannular patch reconstruction of their native pulmonary outflow tract. These data indicate that most of the patients in this previous report had more antegrade pulmonary blood flow and better development of their right ventricular outflow

tracts than the patients included in our report. Nevertheless, their report on the treatment of patients with PS/VSD/MAPCAs is the only previous one that we could identify in the literature.

In summary, the results of this study demonstrate that outcomes for surgical reconstruction of PS/VSD/MAPCAs are excellent. There are two identifiable subgroups defined by the presence or absence of cyanosis during early life. Significant differences between these two groups included the age at first operation, number of procedures to achieve complete repair, right ventricle-to-left ventricle pressure ratios, and late survival. Our institutional experience would suggest that the outcomes achieved for PS/VSD/MAPCAs might be slightly superior to those for PA/VSD/MAPCAs.

## References

1. Kirklin JW, Blackstone EH, Shimazaki Y, et al. Survival, functional status, and reoperations after repair of tetralogy of Fallot with pulmonary atresia. *J Thorac Cardiovasc Surg* 1988;96:102–16.
2. Shimazaki Y, Maehara T, Blackstone EH, Kirklin JW, Barger LM Jr. The structure of the pulmonary circulation in tetralogy of Fallot with pulmonary atresia. A quantitative cineangiographic study. *J Thorac Cardiovasc Surg* 1988;95:1048–58.
3. Rome JJ, Mayer JE, Castaneda AR, Lock JE. Tetralogy of Fallot with pulmonary atresia. Rehabilitation of diminutive pulmonary arteries. *Circulation* 1993;88:1691–8.
4. 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.
5. Carotti A, Albanese SB, Fillepelli 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.
6. Amark KM, Karamlou T, O'Carroll 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.
7. 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.
8. Mumtaz MA, Rosenthal G, Qureshi 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;85:2079–84.
9. DeCampi 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.
10. 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.
11. 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.

12. 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.
13. 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.
14. Kreuzer J, Perry SB, Jonas RA, Mayer JE, Castenada AR, Lock JE. Tetralogy of Fallot with diminutive pulmonary arteries: preoperative pulmonary valve dilation and transcatheter rehabilitation of pulmonary arteries. *J Am Coll Cardiol* 1996;27:1741-7.

---

## The Society of Thoracic Surgeons: Fiftieth Annual Meeting

Mark your calendar for the Fiftieth Annual Meeting of The Society of Thoracic Surgeons (STS) to be held at the Orlando World Center Marriott in Orlando, Florida, January 25-29, 2014. Attend the Annual Meeting to meet the experts, network with colleagues from around the world, participate in a dynamic learning experience, and share an historic moment in the Society's history—its Fiftieth Anniversary.

This preeminent educational event is open to all physicians, residents, fellows, research scientists, perfusionists, physician assistants, nurses, and other interested individuals who work with cardiothoracic surgeons.

Meeting participants will have the opportunity to attend traditional abstract presentations, invited lectures, surgical forums, Early Riser Sessions, Surgical Motion Pictures, and procedural hands-on courses. Parallel sessions on Monday and Tuesday will focus on specific subspecialty interests. The STS Annual Meeting offers more translational science than any other cardiothoracic surgery conference!

An advance program with information about housing and registration will be mailed to STS members this Fall. Nonmembers may contact the Society to receive a copy of the advance program; however, detailed up-to-date meeting information will be available on the STS website at [www.sts.org](http://www.sts.org).

I hope to see you in Orlando.

*Keith S. Naunheim, MD*  
*Secretary*

*The Society of Thoracic Surgeons*  
*633 N Saint Clair St, Ste 2320*  
*Chicago, IL 60611-3658*  
*Telephone: (312) 202-5800*  
*Fax: (312) 202-5801*  
*E-mail: [sts@sts.org](mailto:sts@sts.org)*  
*Website: [www.sts.org](http://www.sts.org)*