Surgical algorithm and results for repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals

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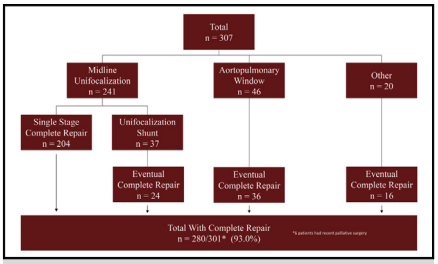
ABSTRACT

Objective: Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries is a complex and heterogeneous form of congenital heart disease. There is a controversy regarding the optimal treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. The purpose of this study was to summarize our algorithm and surgical results for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries.

Methods: This was a retrospective review of 307 patients undergoing primary surgical treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Excluded from this analysis were patients who had undergone prior surgical treatment at another institution and patients with single ventricle and major aortopulmonary collateral arteries. There were 3 surgical pathways, including midline unifocalization (n = 241), creation of an aortopulmonary window (n = 46), and other (n = 20).

Results: For the 241 patients who underwent midline unifocalization, 204 (85.4%) had a single-stage complete repair. There were 37 patients who underwent a midline unifocalization and central shunt, and 24 have subsequently undergone complete repair. Forty-six patients underwent an aortopulmonary window, of whom 36 have subsequently had a complete repair. There were 20 patients who had complex anatomy and underwent procedures other than described, and14 have subsequently undergone complete repair. Thus, for the patients currently eligible, 280 (93.0%) have achieved complete repair. For the 204 patients who had a single-stage complete repair, the mean right ventricle to aortic pressure ratio was 0.36 ± 0.09. Seventy-six patients underwent a staged repair, and the mean right ventricle to aortic pressure ratio was 0.40 ± 0.09 ($P < .05$ compared with single-stage repair). There were 3 (1.5%) early and 8 (4.0%) late deaths for the single-stage complete repair cohort versus 4 (4.0%) early and 15 (14.9%) late deaths for all other procedures ($P < .01$).

Conclusions: The data demonstrate that more than 90% of patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries achieved complete repair. The overall mortality was significantly lower in the subgroup of patients who underwent single-stage complete repair. (J Thorac Cardiovasc Surg 2018;■:1-11)



Flow diagram for the 307 patients with PA/VSD/MAPCAs.

Central Message

This study summarizes our algorithm and surgical results for PA/VSD/MAPCAs. Some 93% of patients underwent complete repair. The mortality was 3.5-fold lower in patients who received single-stage complete repair.

Perspective

PA/VSD/MAPCAs are a complex and heterogeneous form of congenital heart disease. There is an ongoing controversy regarding the optimal treatment of PA/VSD/MAPCAs. This study summarizes our surgical algorithm and results. The data indicate that there were substantial differences in outcome among the several subgroups of patients.

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Abbreviations and Acronyms
MAPCA = major aortopulmonary collateral artery
PA = pulmonary atresia
VSD = ventricular septal defect

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Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs) is a relatively rare and heterogeneous form of congenital heart disease. The natural history of this congenital heart defect is poor, with 10-year and 20-year survivals of 50% and 20%, respectively.[1,2] The early treatment strategies focused on staged thoracotomy procedures and had little impact on prognosis for the group as a whole. On the basis of these results, Bull and colleagues[1] advocated for the abandonment of operations that offered limited benefit to the patient but subjected them to the morbidity of these surgical procedures.

In the mid-1990s, Reddy and colleagues[3] proposed the midline unifocalization approach to PA/VSD/MAPCAs. This approach entails a technically demanding surgical procedure but is extremely versatile in its application. As such, it can be used for the many diverse forms of MAPCA and pulmonary artery anatomy that may be encountered.[4-6] There are numerous groups that have subsequently adopted the midline unifocalization approach and have achieved favorable results compared with the untreated natural history.[7-10]

Despite the relative success of the midline unifocalization approach, there is an ongoing controversy regarding the optimal treatment of PA/VSD/MAPCAs. The Melbourne group has concluded that the unifocalization of MAPCAs confers no long-term benefit, and their group has focused on efforts at ''pulmonary artery rehabilitation.''[11,12] This approach does not include the unifocalization MAPCAs, and they have subsequently reported their results for this approach.[13] There are many other groups that have adopted the pulmonary rehabilitation approach, as evidenced by the plethora of articles that have been published on this subject in the last 10 years.[14-20] It is evident that the optimal surgical treatment of PA/VSD/MAPCAs remains controversial, with the Stanford and Melbourne groups representing the 2 antithetical extremes.

Our group at Stanford has accrued the largest single-center experience in the world with the surgical treatment of PA/VSD/MAPCAs. The purpose of this article is to summarize the Stanford algorithm and experience with the surgical treatment of PA/VSD/MAPCAs.

MATERIALS AND METHODS

This study was approved by the Institutional Review Board at Stanford University (Protocol ID 33924). Patients included in this study had a diagnosis of PA/VSD/MAPCAs and underwent all aspects of their surgical treatment at Stanford. The time frame incorporated begins in 2001 through February 2017. A total of 307 patients met these criteria, including 149 men and 158 women. A total of 209 patients (34%) had DiGeorge syndrome (22q11 deletion), and 13 patients (4%) had Alagille syndrome. The median age at the time of operation was 4.5 months, with a range of 0.1 to 11.5 years (Figure 1, *A*). The median weight at the time of surgery was 5.1 kg (range, 2.1-41.8 kg).

Specifically excluded from this analysis are 167 patients who had surgery performed at an outside institution and were subsequently referred to Stanford for reoperation, because this cumulative experience has been recently published.[21] There were 50 patients who were excluded because they had intracardiac diagnoses other than PA/VSD.[22] Finally, there were 6 patients who required emergency surgical intervention for life-threatening cyanosis and were excluded from analysis. All 6 of these patients died in the short-term or midterm and underscore the futility of such emergency procedures for patients in extremus.

All patients underwent a preoperative cardiac catheterization to delineate the pulmonary artery and MAPCA anatomy. There were 211 patients who were discharged from the hospital after their neonatal cardiac catheterization and subsequently readmitted for repeat cardiac catheterization and elective unifocalization.

A total of 261 patients had predominantly single-supply MAPCAs, as defined by a MAPCA that provides the only source of blood flow to a pulmonary segment. Twenty-nine of the 261 patients had a ductus arteriosus or hemi-truncus to 1 lung and MAPCAs to the contralateral lung. These 29 patients all underwent early surgical intervention because of the presence of high pressure and flow to the lung supplied by the ductus or hemi-truncus.

Twenty-one patients had clinical heart failure not amenable to medical management and therefore underwent early surgical treatment. Forty-six patients (15%) had predominantly dual-supply MAPCAs (defined by $\geq$15 lung segments with dual supply from both the native pulmonary artery system and the MAPCAs) and met our criteria for creation of an aortopulmonary window. These specific criteria also include hypoplastic but confluent and normally arborizing pulmonary arteries in the presence of cyanosis.[23]

The patients had a mean of 4.0 $\pm$ 1.2 MAPCAs. This included 2.2 $\pm$ 1.0 MAPCAs to the right lung (range, 1-5) and 1.8 $\pm$ 0.8 MAPCAs to the left lung (range, 1-4). These data are shown in Figure 1, *B*. A transthoracic pressure line was placed in all patients who underwent a complete repair, and the right ventricle to aortic peak systolic pressure ratios are the values dictated in the operative report at the conclusion of that operation before chest closure.

Results are reported as means $\pm$ standard errors and medians and ranges where appropriate. Statistical analysis to compare groups was performed using Pearson's chi-square or Fisher exact test. For this article, ''operative'' refers to those deaths after the index (initial) operation for each patient, ''interim'' refers to those deaths that occurred after discharge from the index operation but sometime before complete repair, and ''late'' refers to those deaths after complete repair. Comparison of actuarial curves was performed using the log-rank (Mantel–Cox) test (SPSS Statistics version 20.0; SSPS Inc, Chicago, Ill).