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

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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.**

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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

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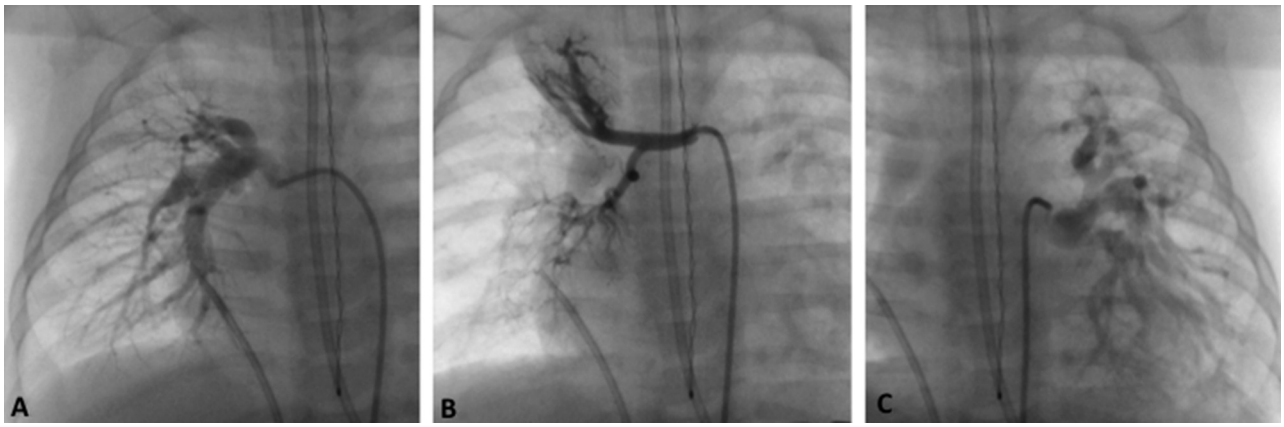

Fig 1. Preoperative angiogram of a typical patient with pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (PA/VSD/MAPCAs) and absence of intrapericardial central pulmonary arteries. The angiogram demonstrates two MAPCAs to the right lung (A, B) and one large MAPCA 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