Pulmonary flow study predicts survival in pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries

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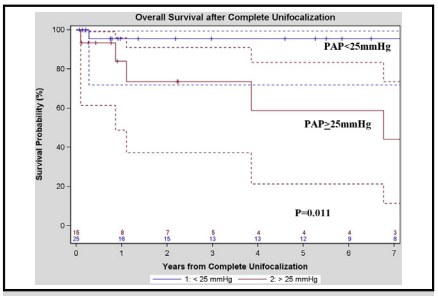
ABSTRACT

Background: We hypothesized that mean pulmonary artery pressure (PAP) detected on a pulmonary flow study may predict medium-term survival and right ventricular systolic pressure (RVSP) in patients with pulmonary atresia (PA), ventricular septal defect (VSD), and major aortopulmonary collateral arteries (MAPCAs).

Methods: Fifty patients with PA/VSD/MAPCAs underwent unifocalization between 2000 and 2013, and 40 of these patients had a pulmonary flow study since 2003. Predictability of the mean PAP on VSD status, medium-term survival, reintervention, and RVSP were analyzed.

Results: Forty-seven of the 50 patients (94%) had complete unifocalization at a median age of 11 months (range, 1-194 months), and 37 patients (74%) achieved VSD closure. Among the 40 patients who underwent a pulmonary flow study, the VSD was closed in 34 (85%), with salvage VSD fenestration in 4 (10%), and was intentionally left open in 6 (15%). Survival was 85.5% at 1 year and 78.5% at 5 years. A mean PAP $\geq$25 mm Hg was associated with worse survival ($P = .011$). Cox regression analysis identified a mean PAP $\geq$25 mm Hg as the sole predictor for death ($P = .037$). Patients with an open VSD had an increased risk of reoperation ($P = .001$) and pulmonary artery reintervention ($P = .010$), and had a trend toward increased risk of death ($P = .059$), compared with those with a closed VSD.

Conclusions: PAP obtained from the intraoperative pulmonary flow study is associated with medium-term survival and late RVSP in patients with PA/VSD/MAPCAs. VSD closure for patients with a mean PAP $\geq$25 mm Hg on a flow study is considered high risk, and sensible judgment and a low threshold for VSD fenestration are required. (J Thorac Cardiovasc Surg 2016;■:1-10)



A high mean PAP ($\geq$25 mm Hg) on a flow study is associated with poor medium-term survival.

Central Message

Mean PAP predicts medium-term survival and RVSP. VSD closure in patients with PAP $\geq$25 mm Hg should be performed with caution.

Perspective

Mean PAP on a pulmonary flow study is a strong predictor for medium-term survival and RVSP regardless of VSD status. VSD closure in patients with a mean PAP $\geq$25 mm Hg is considered high risk, and thus sensible judgment and a low threshold for VSD fenestration are required.

CONG

Pulmonary atresia (PA), ventricular septal defect (VSD), and major aortopulmonary collateral arteries (MAPCAs) pose significant surgical challenges because of the anatomic variations in the native central pulmonary artery and MAPCAs. The ultimate goal of treatment is to achieve a

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Abbreviations and Acronyms

LVSP = left ventricular systolic pressure
MAPCA = major aortopulmonary collateral artery
PA = pulmonary atresia
PAP = pulmonary artery pressure
RV = right ventricular
RVSP = right ventricular systolic pressure
TNPAI = total neo-pulmonary artery index
VSD = ventricular septal defect

biventricular in-series circulation by closing the VSD and establishing an unobstructed pathway from the right ventricle to the pulmonary vasculature with low right ventricular systolic pressure (RVSP). The unifocalization procedure for MAPCAs is a means of incorporating all available lung segments so that the VSD can be closed without significant right ventricular (RV) hypertension. Complete unifocalization has been classically achieved in a staged manner[1-5] and more recently with a single-stage approach,[6-12] with VSD closure rates ranging from 56% to 90%. Nonetheless, the decision to perform VSD closure in patients with borderline PA and MAPCA anatomy always poses significant challenges, and the improper decision for VSD closure may be associated with high mortality and morbidity.

Prediction of the ability to close a VSD was initially attempted using an anatomic index suggested by Reddy et al,[13] the total neo-pulmonary artery index (TNPAI). The TNPAI was found to have an acceptable but suboptimal predictability of VSD closure because of the inability to take stenosis or kinking of MAPCAs and segmental pulmonary vascular resistance into account.[13,14] Reddy et al subsequently proposed using an intraoperative pulmonary flow study as a physiological parameter to reflect total pulmonary vascular cross-sectional area and resistance after complete unifocalization.[13-15] In a previous study, we showed that mean pulmonary artery pressure (PAP) on the intraoperative pulmonary flow study had better accuracy in predicting successful VSD closure compared with TNPAI or other anatomic indices.[16] Nonetheless, the association between mean PAP on a pulmonary flow study and medium-term survival and RV status remain largely unknown. We hypothesized that the postoperative pulmonary vascular status and resultant RV pressure and function may be reflected by the mean PAP on a pulmonary flow study, which potentially predicts medium-term clinical outcomes of this entity.

METHODS

This retrospective study was approved by Research Ethics Board at The Hospital for Sick Children. Fifty consecutive patients with PA/VSD/MAPCAs who underwent at least one unifocalization procedure between January 2000 and December 2013 were identified from the surgical database. The follow-up data were collected up to October 2014. Patients with MAPCAs related to pulmonary stenosis, double-outlet right ventricle, or single-ventricle physiology were excluded. Patient profiles are shown in Table E1.

Surgical Strategy and Technique

The surgical management strategy was consistent throughout the study period.[16] All patients were assessed in view of primary one-stage complete unifocalization via sternotomy at age 4 to 8 months. The staged repair was indicated in 3 circumstances: (1) presence of diminutive native branch pulmonary artery <2 mm, (2) presence of distal stenosis of MAPCAs inaccessible via the sternotomy approach, and (3) critical cyanosis in the neonatal period. A central shunt between the native main pulmonary artery and ascending aorta[17] is typically performed in the neonatal period to rehabilitate the native pulmonary artery or to treat critical cyanosis (arterial saturation <75%). Unilateral unifocalization to a pericardial roll and a shunt (typically 4 mm) is performed via a thoracotomy in patients with MAPCAs with distal stenosis, typically at age 2 to 3 months, followed by contralateral unifocalization or complete repair, depending on the contralateral MAPCA anatomy. Complete unifocalization after those palliative procedures is typically performed by age 8 months.

The surgical techniques for unifocalization and central pulmonary artery reconstruction and the pulmonary flow study are described in detail elsewhere.[16] In brief, our strategy was to unifocalize all important MAPCAs, even those with intraparenchymal communication with other MAPCAs or the native pulmonary artery. MAPCAs were dissected beyond the area of stenosis and then anastomosed to the reconstructed central pulmonary artery without patch materials. The central pulmonary artery was reconstructed with a glutaraldehyde-treated autologous pericardial patch. A homograft patch was not used in this series. The pulmonary flow infusion was incrementally increased from 1 to 2.5 L/min/m^2 through a cannula placed in the reconstructed pulmonary artery. A mean PAP of 2.5 L/min/m^2 was used to determine the adequacy of VSD closure. In recent years, this was increased to 3 L/min/m^2. The cutoff value for VSD closure during the study period was 30 mm Hg, but a more conservative approach was taken in recent years for patients with a borderline mean PAP of 25 to 30 mm Hg who had suboptimal MAPCAs and native pulmonary artery anatomy. The RVSP was measured after VSD closure. The systemic blood pressure from a peripheral arterial line was measured and considered as left ventricular systolic pressure (LVSP). The VSD patch was fenestrated when the RVSP/LVSP ratio exceeded 0.8. The fenestration was created in the middle of the VSD patch, which needed to be at least 5 mm in diameter.

Data Collection

All postoperative echocardiography reports were reviewed to assess RV function. RV dysfunction was defined when qualitatively moderate or severe RV dysfunction was noted on more than 2 consecutive echocardiograms. All preoperative angiographic images were reviewed by an investigator who was blinded to any clinical information. The TNPAI was calculated based on angiography performed just before complete unifocalization using the previously described formula.[13] The follow-up RVSP and RVSP/LVSP ratio were obtained from the latest postoperative cardiac catheterization. The estimated RVSP from echocardiograms was not used. Any catheter-based interventions to the neo-pulmonary artery system, including both unifocalized MAPCAs and native pulmonary artery, and any reoperation after complete unifocalization were documented.

Statistical Analysis

Demographic, anatomic, clinical, and operative characteristics were summarized. Categorical variables are reported as frequency and percentage and compared using Fisher's exact test or the χ^2 test, as appropriate. Continuous variables are presented as mean with standard