Outcomes of surgery for young children with multivessel pulmonary vein stenosis

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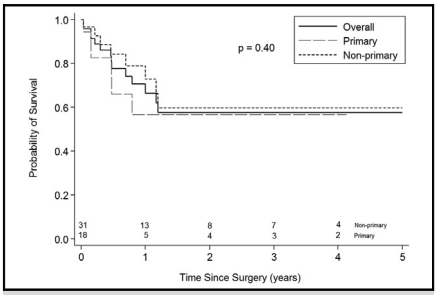
ABSTRACT

Objective: We pursued a multimodality approach to the treatment of patients with pulmonary vein stenosis, incorporating sutureless surgical repair, catheter interventions, and adjunctive chemotherapy. We report our outcomes after surgery.

Methods: Between January 2007 and August 2013, 49 patients with multivessel pulmonary vein stenosis underwent operations at our institution. We retrospectively reviewed data from a pulmonary vein stenosis registry and the medical records.

Results: At the time of the index operation, the median patient age was 6 months (range, 32 days-48 months) and weight was 4.9 kg (range, 2.1-13.4 kg). Fourteen patients (28%) died during the follow-up period (median follow-up was 0.5 years [range, 0.04-4.9 years]). There were 2 deaths (4%) within 30 days. Age at repair <6 months, weight at repair <3 kg, and a preoperative right ventricular systolic pressure < ¾ systemic were found to be associated with mortality. One patient required repeat operation for recurrent stenosis. Thirty-nine patients (80%) underwent postoperative catheterizations. The median number of catheterizations per patient was 2 (range, 0-14). Twenty-nine patients (59%) underwent catheterizations with pulmonary vein intervention. The median number of catheterizations with intervention per patient was 1 (range, 0-14). There were no identifiable associations with need for or number of catheterizations with intervention. Ten patients were listed for lung transplantation: 4 patients were de-listed, 3 patients died waiting, and 3 patients underwent transplant.

Conclusions: Using a multimodality approach, we observed acceptable early survival after operation in patients with pulmonary vein stenosis, despite the need for catheter reinterventions. Lung transplantation remains a viable option. (J Thorac Cardiovasc Surg 2015;150:911-7)



Kaplan-Meier survival curve of patients undergoing operation for multivessel pulmonary vein stenosis.

Central Message

Children undergoing operation for multivessel PVS have acceptable early survival. Catheter-based reinterventions are common but decrease over time.

Perspective

PVS is a challenging disease and survival is poor. We use a multimodality approach, including aggressive surgical resection. With our approach we have achieved acceptable survival. Many patients underwent multiple catheter-based interventions; the frequency decreased with time. Some patients were maintained with ongoing interventions. Surveillance and aggressive treatment of recurrent stenoses is important.

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Pulmonary vein stenosis (PVS) is a disease of neointimal proliferation of myofibroblasts leading to progressive luminal stenosis and obliteration.[1] It may occur in isolation or may be associated with congenital heart disease.[2-5] Catheter-based interventions do not provide effective long-term relief of stenoses.[6,7] The survival of patients with PVS has been poor. Surgical interventions to relieve PVS have included unroofing, patching, and sutureless reconstructions.[8] Surgical series have been limited by small numbers of patients and the results have been generally poor.[9-15] Lung transplantation has also been reported as a viable treatment.[16] Adjunctive chemotherapy has been used to improve the outcome of patients with a poor prognosis.[17] We report our surgical outcomes of infants with multivessel PVS, using a multimodality approach.

METHODS

Institutional review board approval was obtained for this study. Between January 2007 and August 2013, patients with a diagnosis of multivessel PVS (≥2 veins) were identified by review of medical records and a Boston Children's Hospital prospectively collected registry of patients with PVS.

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Read at the 94th Annual Meeting of The American Association for Thoracic Surgery, Toronto, Ontario, Canada, April 26-30, 2014.

Received for publication Aug 11, 2014; revisions received May 6, 2015; accepted for publication June 14, 2015; available ahead of print July 26, 2015.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2015.06.050

Abbreviations and Acronyms
CA = circulatory arrest
CPB = cardiopulmonary bypass
PVS = pulmonary vein stenosis
RV = right ventricle
RSVP = right ventricular systolic pressure

Patient characteristics and all available follow-up data were retrospectively collected. The main objective of this study was to document survival after PVS operation at our institution and need for reinterventions for the overall cohort. The secondary objective was to identify risk factors for these outcomes.

Preoperative Imaging

All patients underwent a transthoracic echocardiogram to assess the status of the pulmonary veins, estimate right ventricle (RV) pressure and function, and identify any other cardiac lesions. All patients had cardiac catheterization to further delineate pulmonary venous anatomy and RV pressure. For this study, the last catheterization before pulmonary vein operation at our institution was used to determine the RV systolic pressure (RVSP) and systemic pressure at rest conditions. If deemed appropriate, PVSs were dilated to decrease pulmonary hypertension. Differential lung perfusion scan, computed tomography, and magnetic resonance imaging were used selectively as adjuvant imaging.

Surgical Technique

All patients had a median sternotomy and hypothermic (18°C) cardiopulmonary bypass (CPB) with cardioplegic arrest. The right atrium was generously opened and the septum primum was resected for optimal intracardiac visualization and assessment of the pulmonary vein orifices. Generally, the approach to the right veins was through Sondengaard's groove. The left veins were approached by retracting the myocardial mass superiorly and toward the midline. Using intermittent periods of circulatory arrest (CA), the pulmonary veins with stenotic lesions were aggressively unroofed and scar tissue was aggressively debrided extending to first-order, and occasionally second-order, branches within the lung parenchyma. Atretic veins were also unroofed if preoperative evaluation showed an open distal vessel. The pulmomary vein branches were then tacked back to the level of the pericardium using simple absorbable polydioxanone suture. Reconstruction of the left atrial well was done using sutureless technique by suturing the pericardium to left atrium using polypropylene suture material. The interatrial septum was usually closed with a 3-4 mm fenestrated piece of autologous pericardium.

Postoperative Care

Routine postoperative cardiac surgical care was provided. In eligible patients, adjunctive chemotherapy with bevacizumab and imatinib mesylate was given as directed by a research protocol (ClinicalTrials.gov NCT00891527). Inclusion in the chemotherapy protocol was at the discretion of the treating team. Follow-up imaging was obtained at the discretion of the attending cardiologist. By echocardiogram, if there was suspicion of recurrent stenoses of a repaired vein or development of stenosis in an unrepaired vein, and/or if there was associated RV hypertension or dysfunction, then a cardiac catheterization procedure was ordered. A priori, the goal was to treat new and recurrent stenoses with catheter-based interventions as much as possible. The decision to intervene was made by the individual interventionalist, largely based on the angiographic appearance of the vessel, and taking into account the overall clinical status of the patient, hemodynamic parameters, and the extent of disease involvement of other lung lobes. Our bias was to intervene on any affected lobar vessel with the appearance of >50% narrowing in 1 view.

All catheterizations were done with general anesthesia and patients often would recover in the intensive care unit. Transseptal puncture was performed, if necessary, to access the pulmonary veins if pulmonary arterial angiography suggested obstruction. Interventions included balloon angioplasty and stenting. Stent placement was reserved as secondary therapy (ie, after balloon dilation failure) except in cases of perceived kinking or obstructive tortuosity of the venoatrial junction. We generally avoided stent placement in peripheral branches or in lobar branches where there was concern for jailing tributary vessels. In some cases, cutting balloons were used, not specifically to treat refractory lesions, but to create a distal aneurysm to allow stable balloon positioning in diffusely small and noncompliant vessels.

Lung transplantation was considered in patients with progressive disease not amenable to further surgical or catheter interventions.

Statistical Analysis

Categorical variables are summarized as number (percent), and continuous variables as median (range). Characteristics were compared for patients with primary versus nonprimary disease using the Fisher exact test for categorical variables and the Wilcoxon rank sum test for continuous variables. Similar comparisons were made for patients with at most 1 pulmonary vein intervention versus those with 2 or more pulmonary vein interventions. Times from surgery to death and from surgery to first catheterization with pulmonary vein intervention were compared using the log-rank test. Rates of catheterizations with pulmonary vein intervention were estimated as the total number of catheterizations with pulmonary vein intervention divided by the total patient-years of follow-up.

RESULTS

Between January 2007 and August 2013, we identified 49 patients with multivessel PVS who who underwent operation at our institution. Eighteen patients (37%) had primary disease (without associated congenital heart disease) and 31 patients (63%) had nonprimary or secondary disease (with associated congenital heart disease). Table 1 gives the clinical characteristics of the entire cohort and by diagnostic group. The primary group was more likely to be premature (born at < 37 weeks gestational age) and tended to have more genetic anomalies. In the group with associated congenital heart disease (n = 31), 5 patients (16%) had heterotaxy and 7 (23%) had single ventricle physiology. Twenty-four patients (49%) had a history of previous cardiac operations and 13 patients (42%) had a diagnosis of total anomalous pulmonary venous return. Nineteen patients (39%) had prior interventions on their pulmonary veins at other institutions.

The median CPB for the operation was 126 minutes (range, 61-257 minutes). The median crossclamp and CA times were 87 minutes (range, 24-229 minutes) and 50 minutes (range, 0-97 minutes), respectively.

Overall, 14 patients died during the follow-up period; mortality 1 year after surgery was estimated as 33% (95% confidence interval, 20%-52%). There were 6 deaths in the primary group and 8 deaths in the nonprimary group; at 1 year after surgery, mortality was 43% in the primary group and 27% in the nonprimary group ($P = .40$). Figure 1 shows the survival for the entire cohort and by