Surgical repair for primary pulmonary vein stenosis: Single-institution, midterm follow-up

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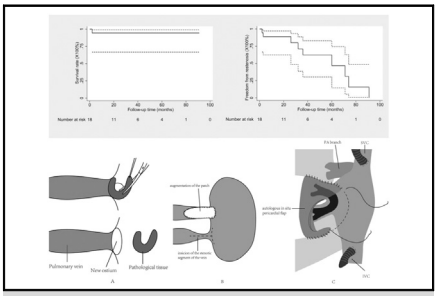
ABSTRACT

Background: Primary pulmonary vein stenosis (PVS) is a rare congenital heart condition and carries a poor prognosis.

Methods: A retrospective review of 18 patients who underwent surgical therapy for primary PVS (2006-2014) was conducted. According to the degree of stenosis severity, the involved pulmonary veins (PVs) were divided into 3 categories: mild (34 veins), moderate (8), and severe (3). Pericardial patch venoplasty was used in 10 involved veins, endarterectomy in 11, and sutureless pericardial marsupialization in 19.

Results: Median surgical age and weight were 19.8 (range: 7-100) months and 7.7 (range: 5.3-20.3) kg, respectively. Bilateral PVS was found in 10 patients (56%), and unilateral in 8 (44%). Moderate or severe stenosis was found more frequently in PVs on the left side ($P = .035$). Multivein involvement was more common in patients age $\leq$18 months than in older patients (75% vs 20%, $P = .054$). No early operative death occurred. Median length of hospital stay was 16 (range: 8-60) days. One subsequent death occurred (6%), at the 2-month follow up after discharge. Median follow-up time for the remaining patients was 29 (range: 2-91) months. Three of the 6 PVs treated for moderate stenosis, compared with 7 of the 34 treated for mild stenosis, developed restenosis, irrespective of the surgical strategy ($P = .153$). Most surviving patients remained in relatively good condition, in New York Heart Association functional class I or II.

Conclusions: Detailed morphologic evaluation of each PV involved is a consideration for surgery, and is closely related to the prognosis. Moderate or severe primary PVS is worse than mild PVS, and no differences were found in effectiveness among endarterectomy, pericardial patch venoplasty, and sutureless pericardial marsupialization in treating primary PVS. (J Thorac Cardiovasc Surg 2015;150:181-8)



This figure shows 3 surgical techniques, and the overall prognosis in the median follow up.

Central Message

Detailed morphologic vein assessment is important for surgery and prognosis in PVS. No differences were found in PVS treatment effectiveness among endarterectomy, patch venoplasty, and the sutureless technique.

Perspective

Use of the sutureless technique as a standard to treat primary pulmonary vein stenosis (PVS), based on effectiveness, is a subject of debate. Our findings indicate a close relationship between morphology and clinical prognosis, with moderate and severe primary PVS being worse than mild PVS. Endarterectomy and pericardial patch venoplasty should be used for treating mild stenosis, and the sutureless technique for moderate and severe PVS.

See Editorial Commentary page 188.

Primary pulmonary vein stenosis (PVS) with normal pulmonary venous connections is a rare congenital cardiac condition (0.4%) and carries a poor prognosis.[1] The clinical presentation can vary from local (discrete and ostial) stenosis, to diffuse stenosis, to atresia. Primary PVS is characterized by obstruction of the pulmonary venous blood flow as a result of hypoplasia of the pulmonary veins (PVs) or of constriction of the intima around the venous-atrial junction, which is caused by abnormal absorption of the common PV into the posterior wall of the left atrium in the later stages of cardiac development.[2]

The condition generally leads to worsening pulmonary hypertension, and severe respiratory and cardiac failure in

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Abbreviations and Acronyms
PV = pulmonary vein
PVS = pulmonary vein stenosis
SPM = sutureless pericardial marsupialization

the pediatric population. Up to 80% of patients have another congenital heart abnormality, such as patent ductus arteriosus, atrial septal defect, and ventricular septal defects.[3,4] These associated conditions elevate pulmonary blood flow and atrial load, which in turn promote PVS.[5] From the histopathologic perspective, neointimal proliferation and medial hypertrophy have been described as main causative pathogenetic processes leading to occlusion of the luminal contour; proliferation of myofibroblast-like cells accounts for this progression.[6-8]

Treatments that include percutaneous and surgical intervention are, unfortunately, followed by progression of the disease, and restenosis within weeks to months, owing to the malignant obliterative disease biology, which is refractory to current therapeutic methods.[9,10] Sutureless pericardial marsupialization (SPM) has been the preferred approach, in many centers, to relieve obstruction and reduce the restenosis rate; its indication has been extended from postrepair PVS to primary PVS.[11-14] However, the results are inconclusive, especially in cases of upstream primary PVS, which usually is not amenable to SPM.[15] The aim of the current study was to review our early and midterm outcomes of surgical correction for this malformation.

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METHODS

Patients

We searched the cardiothoracic database of Shanghai Children's Medical Center, covering records from the 8-year period of October 2006 to February 2014, for patients (n = 18; 7 were males) who presented with primary PVS and were referred to our center for surgical management. Eight patients diagnosed with primary PVS did not undergo surgical repair, owing to parental refusal in 4 cases, and a low severity (stenosis in a single PV) of isolated primary PVS in 3 cases. The other patient (age 6.5 months) unfortunately died in the cardiac care unit before the operation could be performed. These 8 patients were excluded from this study. Institutional review board approval was obtained, and perioperative data were collected and analyzed.

One patient had initial transcatheter closure of patent ductus arteriosus in the local hospital. Primary PVS was defined as anatomically normal pulmonary venous connections without previous surgery. Echocardiography was performed on all patients, and vessels were considered stenotic[16] when the color Doppler technique revealed a turbulent, continuous flow with an abnormally high peak velocity (>1.6 m/s). Angiographic computed tomography is a noninvasive procedure and can rapidly provide excellent images with good spatial resolution that can be used for diagnostic confirmation. Given that the echocardiography results did not reconcile with the computed tomography results in 4 patients, cardiac catheterization was performed.

Pulmonary vein atresia, representing the most severe end of the primary PVS spectrum, was defined either as complete obliteration of the lumen, extending >5 mm within the involved veins on CT or catheterization, or as no demonstrable flow signal on echocardiogram. Table 1 shows the patient characteristics during the study period. Considering a combination of left-to-right-shunt anomaly, which can be associated with the promotion of PVS, and the presence of clinical symptoms, we took an aggressive approach in performing the operation in these patients.

Pulmonary hypertensive crisis was defined as having ≥1 event in which the pulmonary arterial systolic pressure equaled or exceeded systemic levels, followed by a rapid decrease in systemic arterial pressure. Partial relief was defined as alleviating the obstruction, while the velocity of the addressed PVs remained at 1.2 to 1.6 m/s. Restenosis was defined as being either new stenosis after complete relief, progression of the stenosis after partial relief, or total occlusion of the atretic veins that were left unhandled.

Evaluation of Obstructive PVs

Morphologic assessment of the involved veins was made by computed tomography angiography or catheterization. In addition, intraoperative inspection played an important role in the evaluation. Upstream PVs were defined as veins near the pulmonary hilum, or extending into the periphery of the lung; downstream PVs were defined as veins localized proximal to the venoatrial junction, including the ostium. Three types of stenotic PVs were involved in our series. Type-I (mild) was stenosis occurring only at the ostial level or around the venoatrial junction, with a minimum PV diameter of approximately 2 to 4.5 mm. Type-II (moderate) was tubular long-segment or diffuse hypoplasia of the PVs (both downstream and upstream), with a minimum PV diameter of <2 mm. Type-III (severe) was pulmonary vein atresia, such that the atretic veins could not be found, or only a few remnant tissues could be found intraoperatively.

A total of 45 PVs were stenotic. The frequency of involved PVs included the right upper PV in 13 patients, the right inferior PV in 9, the left inferior PV in 12, and the left upper PV in 11. Mild stenosis was found in most of the involved PVs (n = 34), including 12 right upper, 8 right inferior, 7 left inferior, and 7 left upper. A moderate or severe degree of PVS was found in 11 PVs, including 5 left inferior, 4 left upper, 1 right upper, and 1 right inferior, among which atresia was observed in 3—right inferior, right upper, and left inferior. Moderate or severe stenosis of individual PVs was more commonly found in those on the left than those on the right (9 of 23, or 39%, vs 2 of 22, or 9%; $P = .035$).

Ten patients (56%) had bilateral PVS, whereas 8 patients (44%) had unilateral PVS, among whom 3 had stenosis of a single right upper PV. Of the 18 patients, 15 (83%) had mild or moderate stenosis in the involved PVs. We found 6 patients (75%; 6 of 8) in the infant cohort (age ≤18 months), compared with 2 patients (20%; 2 of 10) who were diagnosed in late childhood (age >18 months), who had stenosis of 3 or 4 PVs ($P = .054$). Four (67%) of the 6 infant patients had moderate or severe stenosis.

Surgical Technique

All patients underwent surgical correction for primary PVS and associated defects after standard aorto-bicaval cardiopulmonary bypass was established. The procedures were performed under mild hypothermia, and myocardial protection was provided with cold 4:1 blood cardioplegia administered every 30 minutes during the ischemic time. In most patients, pulmonary venous return can be well controlled with pump suckers. However, facilitating exposure was difficult in 3 cases undergoing SPM, when we put the suturing line near the intraparenchymal portion. In these cases, a temporal reduction of bypass flow was adopted to provide a satisfactory intraoperative visualization.

Endarterectomy was performed in 11 stenotic veins. Exposure of the stenotic orifices of the individual PVs was attained through atriotomy. Two traction sutures placed on the pathologic tissue at the ostium, in the "12 o'clock" and "6 o'clock" directions, can help ease the resection of the tissue (Figure 1, *A*). For obstruction localized proximal to the PV ostia, we aggressively excised the stenotic ring (endocardial or fibrous) up to the tunica adventitia, and directly anastomosed the PV to the atrial endocardium, to ensure an unconstructed neo-orifice.

Pericardial patch venoplasty was used in 10 veins (Figure 1, *B*). In this technique, the patch was anastomosed to the adventitia of the vein.