Total anomalous pulmonary venous connection: Outcome of postoperative pulmonary venous obstruction

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Objective: Pulmonary venous obstruction (PVO) is an important cause of late mortality in total anomalous pulmonary venous connection (TAPVC). We aimed to describe current practices for the management of postoperative PVO and the efficacy of the different interventional procedures.

Methods: We conducted a retrospective international collaborative population-based study involving 19 pediatric cardiac centers in the United Kingdom, Ireland, and Sweden. Patients with TAPVC born between January 1, 1998, and December 31, 2004, were identified. Patients with functionally univentricular circulation or atrial isomerism were excluded. All available data and images were reviewed.

Results: Of 406 patients undergoing repair of TAPVC, 71 (17.5%) had postoperative PVO. The diagnosis was made within 6 months of surgery in 59 (83%) of the 71 patients. In 12, serial imaging documented change in appearance of the pulmonary veins. Good-sized pulmonary veins can progress to diffusely small veins and rarely atresia. Patients presenting after 6 months had less severe disease; all are alive at most recent follow-up. Fifty-six (13.8%) of 406 patients underwent intervention for postoperative PVO: 44 had surgical treatment and 12 had an initial catheter intervention. One half underwent 1 or more reinterventions. Three-year survival for patients with postoperative PVO was 58.7% (95% confidence intervals, 46.2%-69.2%) with a trend that those having a surgical strategy did better ($P = .083$). Risk factors for death included earlier presentation after TAPVC repair, diffusely small pulmonary veins at presentation of postoperative PVO, and an increased number of lung segments affected by obstruction.

Conclusions: Postoperative PVO tends to appear in the first 6 months after TAPVC repair and can be progressive. Early intervention for PVO may be indicated before irreversible secondary changes occur. (J Thorac Cardiovasc Surg 2013;145:1255-62)

CHD

There is ongoing late mortality in patients with total anomalous pulmonary venous connection (TAPVC), frequently associated with postoperative pulmonary venous obstruction (PVO). Studies have shown that postoperative PVO can occur in 5% to 18% of patients.[1-8]

We[9] have previously reported morphology and outcomes of all patients born with TAPVC in the United Kingdom, Ireland, and Sweden over a 7-year period (n = 422). Postoperative PVO was found to be an important risk factor for death, occurring in 71 (17.5%) of 406 patients who underwent repair of TAPVC. Risk factors for development of postoperative PVO were identified[9] and comprised preoperative hypoplastic/stenotic pulmonary veins and absence of a common confluence.[9] This article focuses on the group

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Abbreviations and Acronyms
CI = confidence interval
CT = computed tomography
HR = hazard ratio
MRI = magnetic resonance imaging
PVO = pulmonary venous obstruction
TAPVC = total anomalous pulmonary venous connection

of 71 patients with postoperative PVO describing presentation, morphologic features, intervention, and outcome of the interventions.

Owing to the relative rarity of TAPVC and postoperative PVO, previous studies have had to span a wide range of surgical eras to obtain sufficient patients for analysis. The aim of this study was to examine the prevalence, treatment, and outcome of the cohort of patients with postoperative PVO in a contemporary international population-based study.

METHODS

The UK, Ireland and Sweden Collaborative study of TAPVC is a retrospective population-based study of all children born with this disease between January 1, 1998, and December 31, 2004, in the United Kingdom, Republic of Ireland (hereafter denoted as Ireland), and Sweden.[9] Patients with functionally univentricular circulation or atrial isomerism were excluded. Ethics committee approval was obtained. The methodology has been described previously.[9]

The records of each of the patients undergoing surgical repair of TAPVC were assessed for postoperative PVO. Ninety percent of all postoperative diagnostic angiography, 54% of diagnostic magnetic resonance imaging (MRI)/computed tomography (CT) imaging, and 30% of diagnostic echocardiography performed were directly visualized and reinterpreted by 2 investigators (A.N.S., J.P.) along with all catheterization and operative reports. Pulmonary veins were considered to be obstructed on echocardiography if the pulsed wave pulmonary venous Doppler pattern showed nonphasic flow or velocities greater than 2 m/s.[10] Angiographic diagnosis of PVO was gained either from the levophase of pulmonary arterial injection or from direct injection of contrast into the individual pulmonary veins. Pulmonary veins were considered to be obstructed if the pulmonary vein diameter was reduced by 50% or more from the largest measured dimension. The hemodynamic criterion for PVO was a mean gradient across the stenosis of 4 mm Hg or more.[5] All available pathologic reports were studied.

In this cohort with postoperative PVO, the morphologic features assessed comprised the following:

1. Unilateral or bilateral obstruction at presentation of postoperative PVO
2. Type of obstruction: the pulmonary veins were put into 1 of 2 categories on the basis of qualitative observations—either discrete stenosis with normal-sized pulmonary veins or stenosis with diffusely small pulmonary veins
3. Site of PVO
4. Progression of obstruction
5. Pulmonary venous collateral circulation

The angiogram was reviewed in the frontal projection, and lung zones affected by PVO were documented (upper and lower zones).

Statistical Methods

Risk factors with continuous distribution were expressed as median (minimum-to-maximum) and categorical risk factors as number (percent). Patient survival was described using Kaplan-Meier curves, and Cox proportional hazards modeling was used to test the association between potential risk factors and death. Ninety-five percent confidence intervals (CIs) were quoted (STATA 10; Stata Corporation, College Station, Tex). Multivariable analysis was not possible owing to the relatively small number of patients and variables.

RESULTS

Four hundred twenty-two live births with TAPVC were identified, and 406 of these infants underwent surgical repair; 71 (17.5%) of the 406 had evidence of postoperative PVO.[9] These 71 are the subject of this report. Surgery was performed in multiple institutions by many different surgeons as previously described.[9] A wide variation in surgical technique was noted.

Presentation of Postoperative PVO

PVO was diagnosed 0 days to 5.2 years (median, 49 days) after TAPVC repair, including 4 patients in whom obstruction was identified in the operating room at the time of primary repair. Two of these 4 patients died 4 and 9 days after TAPVC repair and had hypoplastic and stenotic individual veins at autopsy. The other 2 were known to have mild residual obstruction.

In an additional 12 patients, PVO was diagnosed before initial hospital discharge. Ten of them underwent repeat surgery during the same admission for attempted relief of PVO. A total of 65 (92%) of the 71 achieved hospital discharge, with 55 cases of PVO being diagnosed after initial hospital discharge. Among these 65 patients, 8 were relatively asymptomatic with evidence of obstruction being found during routine follow-up; 4 had no intervention inasmuch as the disease was very mild. The others (n = 57) exhibited breathlessness and/or failure to thrive. Overall, 59 (83%) of 71 cases of PVO were diagnosed within 6 months of initial surgery.

Diagnostic Imaging for the Presence of PVO

All patients underwent echocardiography. No further imaging was performed in 21 (30%), including 2 who died before further investigation could be performed. Thirty-eight (54%) had angiography, and 7 also had MRI and/or CT imaging.

Morphology

Of the 71 patients, 25 had supracardiac, 25 infracardiac, 11 mixed, and 9 cardiac site of connection. There was also 1 with common pulmonary vein atresia. This represents 25 (12%) of 205 in the overall cohort with supracardiac TAPVC, 25 (23%) of 110 in the cohort with infracardiac TAPVC, 11 (30%) of 37 in the cohort with mixed TAPVC, and 9 (13%) of 67 in the cohort with cardiac TAPVC.