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)

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.

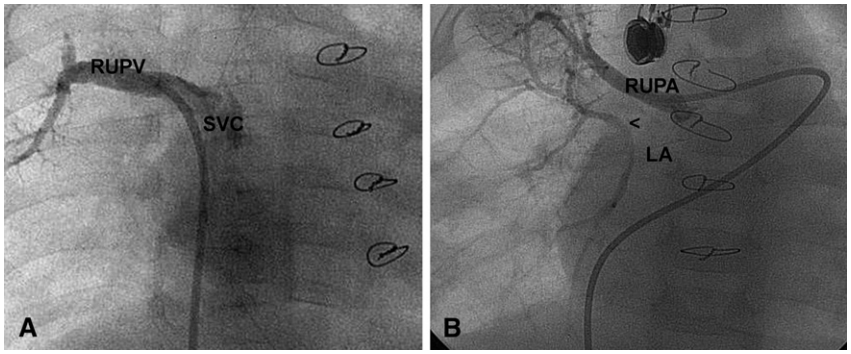

FIGURE 1. Example of progressive change in the size of the right upper pulmonary vein *(RUPV)* in a patient with mixed total anomalous pulmonary venous connection *(TAPVC)*. (A) Before TAPVC repair, a good-sized RUPV connects to the proximal superior vena cava (*SVC*). (B) After TAPVC repair and attempted surgical relief of RUPV obstruction, the RUPV appears to be diffusely small (<) after injection of contrast into the right upper pulmonary artery (*RUPA*). Time between angiograms was 2.4 years. *LA*, Left atrium.

At angiography, it was often difficult to differentiate clearly whether a discrete obstruction was at the anastomosis or separate and upstream from the anastomosis at the pulmonary venous ostium. We therefore used the operation notes to help localize the site of obstruction; however, these only described the anatomy at the time of surgical intervention and not at initial presentation. We had data on 50 of 52 children having surgery at some point in their treatment for postoperative PVO. In most cases the morphologic features were complex, with narrowing at the anastomosis between the confluence and the left atrium resulting in retraction and fibrosis of the surrounding tissues, which produced narrowing at the entrance of the pulmonary veins. In rare cases (n = 6), the surgical report stated that the stenosis was at the pulmonary vein ostium with the anastomosis being unaffected.

Fifty-four patients had bilateral and 15 had unilateral PVO: 8 on the right and 7 on the left. In 2, the pattern of stenosis was unclear. At presentation, 35 had discrete stenosis with normal-sized pulmonary veins, 14 had stenosis with ''small'' pulmonary veins as defined in the Methods section (9 of whom had small pulmonary veins before TAPVC repair), and in 22 the presenting morphologic features were not known. In 14, one or more of the pulmonary veins was atretic.

In 19 (26.8%) of 71 patients, there were 2 or more angiographic, CT, or MRI studies. In 12 of the 19, serial imaging could document changes in the appearance of the pulmonary veins over time. Time between studies where changes were seen varied from 20 days to 2.4 years. Figure 1 illustrates how good-sized pulmonary veins progressed to diffusely small pulmonary veins and, rarely, atresia after obstruction. If unilateral, the pulmonary artery became small on the same side. One patient progressed from unilateral to bilateral disease. In 16 patients, venovenous collaterals of varying sizes formed, draining to the systemic veins or other lesser-affected parts of the pulmonary venous bed. In half, the collateral system formed extensive networks; in 1 case a pulmonary venous collateral crossed the midline to the unaffected lung.

INTERVENTION FOR POSTOPERATIVE PVO

Sixty (14.8%) of the 406 undergoing surgical repair of TAPVC had postoperative PVO requiring intervention. Four patients either died before intervention could be performed or were awaiting intervention at last follow-up; therefore, 56 (13.8%) of 406 underwent intervention for postoperative PVO. Half (28/56) required more than 1 intervention.

This was an observational study, and the choice of intervention was made by the treating physician. Forty-four had

TABLE 1. Initial intervention for relief of postoperative PVO (n = 56)

Procedure	No.	No. alive
Conventional balloon angioplasty	11	7
Cutting balloon angioplasty	1	1
Anastomotic/PV enlargement with pericardial patch	12	3
Anastomotic/PV enlargement with PTFE	3	1
Anastomotic/PV enlargement with native LA tissue	8	5
PV plasty/dilatation/endarterectomy	6	5
Redo coronary sinus unroofing	2	2
Right PVs/anastomotic enlargement with sutureless pericardial patch. Left PV enlargement with LA appendage	1	1
Right PVs/anastomotic enlargement with sutureless pericardial patch	1	1
Enlargement of the PV channel by suturing veins together	2	1
Excision of fibrous ridge	1	0
Excision of stenosis, redo of shrunken ASD patch	1	1
Enlargement of anastomosis by extending the incision	3	3
Redo homograft patch diverting blood into LA and enlargement of ASD	1	1
Unknown surgical details	3	1

PVO, Pulmonary venous obstruction; *PV*, pulmonary vein; *PTFE*, polytetrafluoroethylene (Gore-Tex; W. L. Gore & Associates, Inc, Flagstaff, Ariz); *LA*, left atrium (atrial); *ASD*, atrial septal defect.

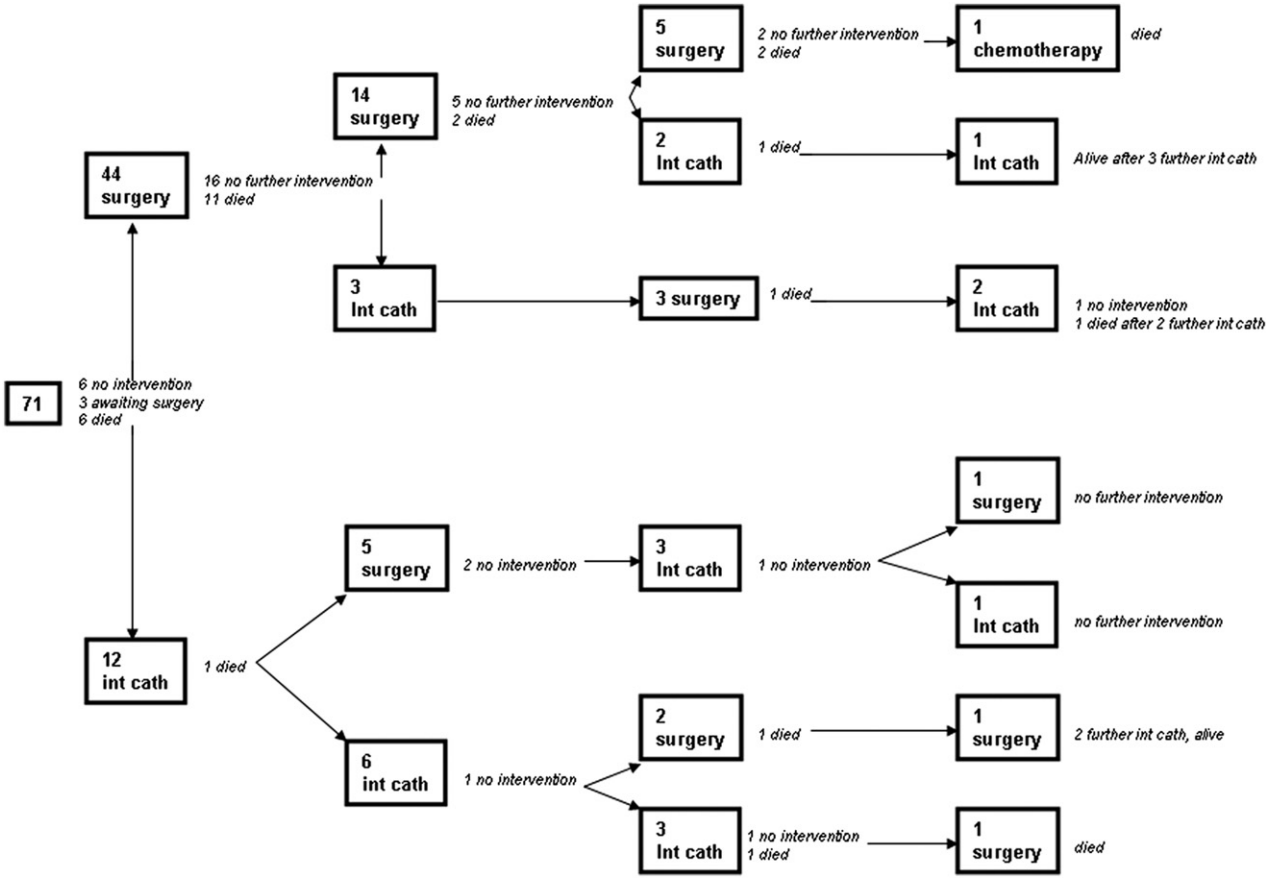

FIGURE 2. Interventions performed for postoperative pulmonary venous obstruction. *Int cath*, Interventional catheter.

an initial surgical intervention and 12 an initial catheter intervention. Table 1 details the initial interventions performed and Figure 2 outlines further procedures performed. Patients having an initial catheter intervention had a higher pulmonary artery/systemic blood pressure ratio, but otherwise there were no significant morphologic differences between the catheter and surgical groups (Table 2).

Many different types of surgical intervention were performed, making analysis of individual procedures impossible. Altogether, 75 surgical procedures of various types were performed on 52 patients, 10 using the sutureless technique (on 9 patients). The sutureless technique was only used in 2 patients as the initial intervention for postoperative PVO.

TABLE 2. Clinical comparison of patients undergoing *initial* catheter (n = 12) or surgical (n = 44) intervention for postoperative PVO

	Catheter intervention	Surgical intervention	*P* value	No.*
Pulmonary artery/systemic blood pressure ratio, median (IQR)	0.75 (0.50, 0.89)	0.50 (0.44, 0.63)	.049	16
Discrete stenosis of 1 or more pulmonary veins	6 (54.6%)	23 (76.7%)	.168	41
Lung segments involved				54
1	0 (0%)	3 (7.1%)	.401	
2	4 (33.3%)	6 (14.3%)		
3	1 (8.3%)	3 (7.1%)		
4	7 (58.3%)	30 (71.4%)		
Unilateral disease	3 (25.0%)	8 (18.6%)	.624	55
No collaterals	7 (58.3%)	11 (55.0%)	.622	32
Minor collaterals	2 (16.7%)	6 (30.0%)		
Major collaterals	3 (25.0%)	3 (15.0%)		
Age (d) at intervention (median, IQR)	44 (28,69)	47 (38,74)	.632	56

PVO, Pulmonary venous obstruction; *IQR*, interquartile range. *Number of patients in whom data were available.

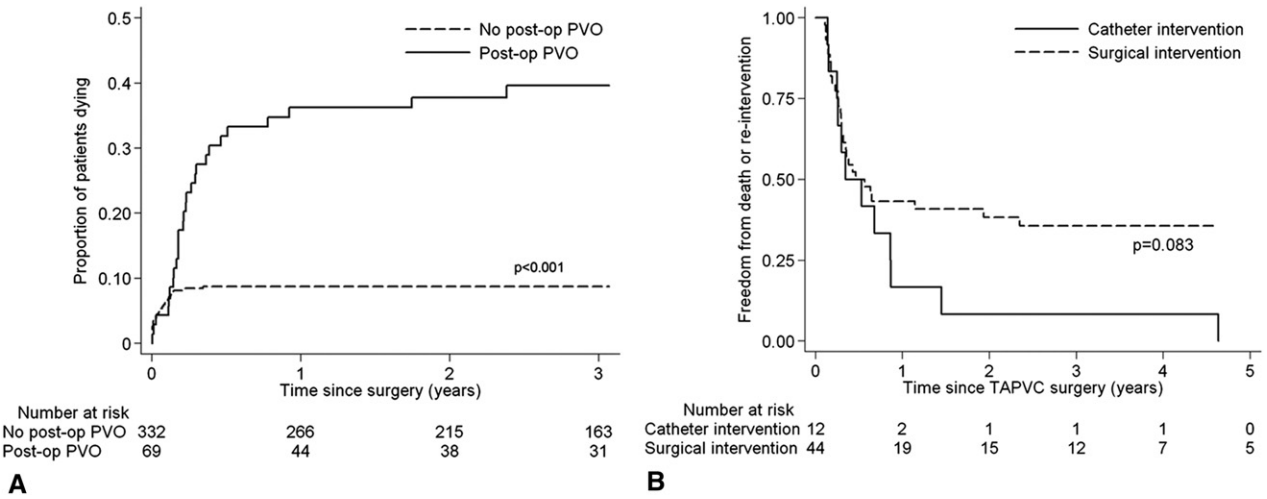

FIGURE 3. Kaplan-Meier curves showing (A) survival of patients having surgical repair of total anomalous pulmonary venous connection *(TAPVC)*, with and without postoperative pulmonary venous obstruction *(PVO)*, and (B) freedom from death or second reintervention for those having an initial surgical and catheter intervention for postoperative PVO. Patients were censored in part B if they had a further catheter or surgical intervention or died. Of the interventional catheter group, all underwent reintervention (n = 11) or died (n = 1) within a year of their first intervention for postoperative PVO. *Time zero* is the date of initial TAPVC repair.

CHD

Forty catheter interventions were performed on 17 patients: 4 cutting-balloon procedures (on 2 patients) and 4 stenting procedures (on 3 patients); the remainder were conventional balloon dilatations. There was 1 hybrid procedure performed.

Outcomes for Postoperative PVO

Median follow-up of all patients undergoing surgical repair of TAPVC (n = 406) was 2.9 years (range, 1 day to 8.4 years). Two patients died of causes unrelated to the heart. Only 6 patients died more than 6 months after TAPVC repair, all secondary to postoperative PVO. There were no deaths after 3 years.[9]

All patients with PVO. Median follow-up for cases of postoperative PVO was 2.3 years (range, 1 day to 8.1 years) representing 192.0 person-years of follow-up; in that time there were 29 deaths.

TABLE 3. Risk factors for (1) death in univariable analysis of the 71 patients in whom postoperative PVO was diagnosed and (2) death or reintervention in univariable analysis of the 56 patients who had an intervention for postoperative PVO

	Seventy-one patients with postop PVO				Fifty-six patients that had an intervention for postop PVO			
Variable	HR	95% CI	*P* value	No.*	HR	95% CI	*P* value	No.*
Diffusely small pulmonary veins	6.54	2.5-17.07	<.001	49	2.40	1.18-4.88	.016	41
Age at presentation with postop PVO (wk)†	0.87	0.81-0.93	.001	71	0.99	0.97-1.00	.109	56
Time from initial repair to diagnosis of postop PVO (wk)†	0.78	0.69-0.88	<.001	71	0.99	0.97-1.00	.109	56
No. of lung segments involved	1.70	1.03-2.82	.038	69	1.13	0.81-1.58	.470	54
No collaterals	1.00	Reference		38	1.00	Reference		32
Minor collaterals	0.85	0.17-4.19	.837	38	3.63	1.35-9.74	.010	32
Major collaterals	2.94	0.88-9.77	.079	38	1.03	0.37-2.90	.957	32
Pulmonary artery/systemic blood pressure ratio‡	1.00	0.77-1.30	.982	20	1.05	0.91-1.23	.508	16
Unilateral disease	0.34	0.10-1.13	.078	70	0.74	0.33-1.67	.466	55
Cardiac TAPVC	1.00	Reference		71	1.00	Reference		56
Infracardiac TAPVC	1.06	0.29-3.91	.931	71	0.97	0.28-3.37	.957	56
Mixed TAPVC	2.34	0.61-9.08	.217	71	1.46	0.38-5.67	.581	56
Supracardiac TAPVC	1.04	0.28-3.85	.952	71	1.60	0.46-5.49	.458	56
Obstruction before initial TAPVC repair	0.99	0.46-2.15	.984	68	1.43	0.72-2.83	.308	54

PVO, Pulmonary venous obstruction; *HR*, hazard ratio; *CI*, confidence interval; *TAPVC*, total anomalous pulmonary venous connection. *Number of patients in whom data were available. †These risk factors are extremely similar/overlapping: both are included for interest. ‡Data gained from either diagnostic cardiac catheterization before surgical intervention or during interventional catheter procedure.

Estimates of 30-day, 1-year, and 3-year survival for all patients with postoperative PVO were 95.8% (95% CI, 87.5%-98.6%), 62.0% (49.7%-72.1%), and 58.7% (46.2%-69.2%), respectively.[9] This is significantly worse (hazard ratio [HR] = 4.70; $P < .001$; 95% CI, 2.80-7.87) than that of patients without postoperative PVO, in whom 30-day, 1-year, and 3-year survivals were 94.0% (90.8%-96.1%), 91.2% (87.6%-93.8%), and 91.2% (87.6%-93.8%), respectively (Figure 3, *A*).

Analysis of the 71 patients with a diagnosis of postoperative PVO showed that risk factors for death comprised diffusely small pulmonary veins (HR, 6.5; 95% CI, 2.5-17.1; $P < .001$) and an increased number of lung segments affected (HR, 1.70; 95% CI, 1.03-2.82; $P = .038$) (Table 3).

The time between initial TAPVC repair and presentation with postoperative PVO was also important; patients presenting later with postoperative PVO did better (HR, 0.78; 95% CI, 0.69-0.88; $P < .001$). None of the patients in whom postoperative PVO appeared later than 6 months after TAPVC repair died. All had discrete obstruction rather than diffusely small pulmonary veins at presentation. Two required repeat interventions owing to restenosis but both survived; the disease process of PVO was less severe in the late presenters. Age at presentation with postoperative PVO is a similar/overlapping variable and not surprisingly was important, with older patients presenting with postoperative PVO doing better (HR, 0.87; 95% CI, 0.81-0.93; $P = .001$).

Patients with PVO undergoing intervention. Among the patients undergoing intervention for postoperative PVO (n = 56), there appears to be a trend for the surgical group to fare better than the catheter group with better freedom from death or reintervention; however, this was not statistically significant (HR, 0.55; CI, 0.28-1.08; $P = .083$) (Figure 3, *B*). In these 56, patients with diffusely small pulmonary veins at diagnosis of postoperative PVO did worse than those with discrete stenosis (HR, 2.40; 95% CI, 1.18-4.88; $P = .016$) (Table 3). When collaterals were present, patients with a poorly formed collateral system did worse (HR, 3.63; 95% CI, 1.35-9.74; $P < .010$).

DISCUSSION

The UK, Ireland and Sweden Collaborative study of TAPVC differs from other studies of this disease because it is international, multicenter, and population-based, enabling a relatively large number of patients to be identified within a short time period. This gives insight into contemporary management of the disease in contrast to other studies, which frequently study patients over several decades to gather sufficient patients.

Incidence of Postoperative PVO

We found a relatively high incidence of PVO compared with other series reported recently.[6] This is likely to be a result of our methodology. Because our study is population- rather than intervention-based, we captured all, or very nearly all, patients with postoperative PVO. This includes those who died before intervention and those with mild obstruction insufficient to warrant intervention.

Diagnosis of Postoperative PVO

In 83% of the patients with postoperative PVO, the diagnosis was made in the first 6 months after TAPVC surgery. Typically, a continuous-flow pattern on pulsed wave Doppler, rather than the maximum velocity of pulmonary venous flow, alerted the clinician to obstruction. With postoperative TAPVC, all the flow goes through 1 orifice and the flow pattern is dependent on its relative size. Echocardiography alone may not diagnose and describe the anatomy adequately, particularly where only 1 vein is involved.[10] Conversely, there were several examples in which PVO was suspected on echocardiography but subsequently excluded by angiography, CT, or MRI. In view of this, we believe that all patients with suspected PVO should have additional imaging to echocardiography to delineate the anatomy and enable early diagnosis of postoperative PVO. It is possible that in this group with postoperative PVO, some mild residual obstruction may have been present immediately after initial repair of TAPVC but not recognized. With recent improvements in noninvasive imaging technology, visualization of the pulmonary veins has improved greatly, especially with high-resolution CT angiography and MRI.[11]

Morphology and Progression

Our previous work showed the significant morphologic spectrum found in TAPVC.[9] Our previous publication has shown that preoperative features do have a role in the development of postoperative pulmonary vein stenosis.[9] In particular, multivariable risk factors for postoperative PVO comprised preoperative hypoplastic/stenotic pulmonary veins and absence of a common confluence.[9] Most cases of mixed TAPVC will not have a single confluence, and indeed 30% of the whole cohort with mixed TAPVC developed postoperative pulmonary vein stenosis. Some have speculated that newer surgical techniques such as a primary sutureless repair may help prevent postoperative PVO.[12] Recent reports are encouraging.[13,14]

There appear to be 2 groups of patients with postoperative PVO:

1. Young patients presenting early (<6 months) after TAPVC repair, often within the first few weeks, generally with aggressive and progressive obstruction
2. Older patients presenting later with milder and less progressive disease

In our study, the 12 patients presenting more than 6 months after TAPVC surgery are all alive at latest follow-up.

Caldarone and associates[8] previously found that patients with more diffuse disease of the individual pulmonary veins tended to have more rapid progression and died. Hyde and colleagues[4] similarly found that median time to intervention was shorter in patients with stenosis of the individual veins compared with purely anastomotic obstruction. Ricci and associates[15] also found early presentation of postoperative PVO to be a risk factor for death. The results of our study support these findings. We found that (1) diffusely small pulmonary veins at diagnosis of postoperative obstruction and (2) a shorter time between initial TAPVC repair and presentation with obstruction were both risk factors for death. All but 1 of the patients with diffusely small pulmonary veins presented within 3 months of TAPVC surgery. Of those presenting after 6 months of TAPVC surgery, all had discrete obstruction rather than diffusely small pulmonary veins.

The way the pulmonary veins react to obstruction after TAPVC repair is similar to that seen in pulmonary vein stenosis owing to other causes, suggesting a possible common mechanism. We[16] have previously shown that PVO tends to progress; in some cases venovenous collateral vessels will form, and a major collateral system can be a protective feature.[16] Unfortunately, we had limited access to postmortem data and were unable to study histologic characteristics. The pathologic mechanisms responsible for progression of PVO need to be elucidated. It is possible that some patients have genetic factors that may predispose them to development of stenosis.[17] Further investigation is needed to examine the histologic and biochemical pathways responsible for all forms of PVO; this is likely to require a pathology-based prospective collaborative multicenter study.

Owing to the progressive nature of PVO, early intervention before irreversible secondary changes occur may be indicated. We have shown poor outcome when the pulmonary veins have become diffusely small.

Interventions

Institutions with significant experience with the sutureless technique have recently reported encouraging short- and medium-term outcomes.[18,19] We had anticipated finding more examples of this technique, but only 9 patients had a sutureless repair. This may reflect the time period of our study (1995-2004) compared with when the technique was published (1996 and 1998)[20,21] and reluctance to adopt a new technique in the absence of long-term outcome data.

Many different surgical[20-22] and catheter[23-25] techniques exist to treat PVO. The plethora of interventions and the small numbers in each group made comparative analysis problematic. In addition, as morphologic features of pulmonary vein stenosis varied, surgeons altered technique to account for individual patient's anatomy, making analysis even more difficult. Future studies comparing techniques to treat postoperative PVO should be prospective, consider pulmonary vein morphologic features, and consider severity of the disease. A pulmonary vein score, such as that devised by Yun and colleagues,[18] would help evaluate outcomes of different interventions.

Our data show a trend toward lower reintervention rate in those undergoing an initial surgical rather than catheter approach for postoperative PVO. This was, however, not statistically significant. This is similar to our findings in patients having intervention for primary pulmonary vein stenosis.[16] We should be wary in interpreting these data inasmuch as the underlying morphologic substrate may have been different. For example, an initial catheter strategy may have been chosen for sicker patients with more diffuse stenosis including disease in the individual pulmonary veins. This is supported by our data, which show a statistically higher pulmonary/systemic pressure ratio in those undergoing an initial catheter procedure. Likewise, patients undergoing surgical repair using polytetrafluoroethylene (Gore-Tex; W. L. Gore & Associates, Inc, Flagstaff, Ariz) or bovine pericardium appeared to fare less well, but this may simply reflect patients with more severe disease requiring more extensive reconstruction (Table 1).

Owing to the progressive nature of this disease, the most important factor is whether complete abolition of the stenosis is achieved. Clinicians should strive for this, whichever technique is used.

Study Limitations

Owing to the collaborative and observational design of the study, there was considerable variation in surgical technique. Owing to different patients, centers, and operators, the study design is not optimal for comparing different interventions. In addition, although this is a large study, numbers are still too small to assess the relative advantages of individual interventions clearly. Relatively small numbers also prevented us performing a meaningful multivariable analysis.

CONCLUSIONS

We have identified a cohort of patients in whom PVO developed after surgical repair. PVO after repair of TAPVC remains a difficult and complex disease to treat, with 40% of patients dying within 3 years of TAPVC repair. Postoperative PVO usually develops in the first 6 months after surgical repair and is progressive. Close observation is required during this period, with further imaging being performed if there is any suspicion of postoperative PVO. In younger patients, postoperative PVO is frequently progressive, with pathologic remodeling of the pulmonary venous bed. The mechanisms remain unknown and need further clarification. Early intervention may be indicated before irreversible

changes occur. Patients presenting later than 6 months after TAPVC repair appear to have a less progressive course.

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