
Ujjwal K. Chowdhury*, Diplomate NB, Sukhjeet Singh, Niwin George, Suruchi Hasija, Lakshmikumari Sankhyan, Srikant Sharma, Diplomate NB, Niraj Nirmal Pandey, Mani Kalaivani,
Cardiothoracic Sciences Centre All India Institute of Medical Sciences, New Delhi, India.

*Corresponding author: Ujjwal Kumar Chowdhury, M.Ch., Diplomate NB Professor Department of Cardiothoracic and Vascular Surgery AIIMS, New Delhi-110029, INDIA.

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Abstract

Background: Unligated vertical vein in repaired obstructive totally anomalous pulmonary venous connection (TAPVC) with pulmonary hypertension reduces perioperative pulmonary artery pressure, pulmonary hypertensive crises and improves survival. Our aim was to assess the long-term results of delayed ligation of the vertical vein using an adjustable ligature on survival and reoperations.

Method: A series of 99 consecutive patients (62 males), aged 24.8±55.3 months (range 1 day-25 years; median 4 months) underwent rechanneling of isolated, obstructive and non-obstructive TAPVC with left atrial augmentation, atrial septal fenestration and an unligated vertical vein. An adjustable vertical vein ligature was employed in all patients for later interruption.

Results: Hospital mortality was 5.0% (n=5) with 2 (2%) late deaths. At a mean follow-up of 218.8 (SE± 209.9) months, the actuarial survival was 93.7% (SE± 0.02%; 95% CI: 86.5, 97.1). Postoperatively, all ligatures were tightened gradually over a period of 24-144 hours. Computed-tomographic angiograms during follow-up revealed absence of flow through the vertical vein and ruled out distortion of the left superior pulmonary vein and left brachiocephalic vein.

Conclusions: A patent vertical vein, augmented left atrium and atrial septal fenestration in repaired obstructive and non-obstructive TAPVC with pulmonary hypertension decreased postoperative pulmonary hypertensive crises and improved survival by providing superior hemodynamics. Percutaneously adjustable vertical vein ligature is an expedient, safe and effective technique in these patients in a setting where transcatheter solutions are not available. Use of a percutaneously adjustable ligature around the vertical vein allows gradual tightening of the ligature under optimal physiological conditions, without multiple reoperations (Figure 1).

Keywords: adjustable vertical vein ligature; atrial septal defect; left atrium; cardiac anatomy; congestive cardiac failure; low cardiac output syndrome; obstructive totally anomalous pulmonary venous connection; pulmonary hypertensive crisis; right superior pulmonary vein; systolic pulmonary artery pressure; totally anomalous pulmonary venous connection;

Running title: Adjustable Vertical Vein Ligation

Abbreviations and Acronyms

ASD : Atrial septal defect
CCF : Congestive cardiac failure
CPB : Cardio pulmonary bypass
CPVC : Common pulmonary venous chamber
CVP : Central venous pressure
CT : Computed-tomography
ECMO : Extra corporeal membrane oxygenation
LA : Left atrium
LAP : Left atrial pressure
LCOS : Low cardiac output syndrome
LV : Left ventricle
**Introduction**

Despite fine tuning of surgical techniques and pulmonary vasodilators, recurrent episodes of pulmonary hypertensive crises and low cardiac output remains a significant problem in repaired obstructive TAPVC leading to hemodynamic instability and mortality after surgery [1–4].

Studies have demonstrated that the left atrium lacks both normal compliance and reservoir function and the left ventricle (LV) is non-compliant and dysfunctional in a subset of patients with obstructive TAPVC [5–11]. It is postulated that a patent vertical vein may function as a temporary reservoir for pulmonary venous blood after repair of TAPVC, permit unloading of the volume in the small, non-compliant left-sided cardiac chambers until they are able to grow and adapt to increased flow demands [5, 6, 12, 13]. Not all investigators have accepted these findings or utilized these techniques. Clinical studies on an unligated vertical vein in the setting of obstructive TAPVC are limited and insufficient to generate evidence-based guidelines.

In our previous investigation on ligation versus non ligation, we demonstrated that unligated vertical vein in repaired obstructive TAPVC reduces perioperative pulmonary artery pressure, pulmonary hypertensive crises and improves survival [14]. Subsequently, in 2007, we demonstrated that a percutaneously adjustable vertical vein ligature allows gradual tightening of the ligature under optimal physiologic conditions without reopening the sternum [15]. In the current study, we did not repeat the above analysis.

The primary objective of this study was to i) determine the short-and long-term impact of delayed vertical vein ligation using an adjustable vertical vein ligature on survival of patients with supracardiac TAPVC having moderate or severe pulmonary artery hypertension. The secondary objectives of the study were to: ii) examine whether the unligated vertical vein is serving as a temporary “pop-off” valve in the event of pulmonary hypertensive crises; iii) ascertain the timing to occlude the vertical vein; iv) evaluate the presence of iatrogenic kinking of left superior pulmonary vein and left brachiocephalic vein, if any and v) study the requirement of cardiac reoperations for residual shunts at the level of vertical vein or recurrent pulmonary venous obstruction.

**Methods**

This retrospective study conforms to the principles outlined in the declaration of Helsinki and was approved by the Institutional Ethics Committee. Patients were enrolled in the study protocol after obtaining informed written consent from patients/parents/guardians.

**Patient selection criteria**

This study evaluated short- and long-term outcomes after delayed ligation of the vertical vein in a consecutive series of patients undergoing repair of isolated supracardiac TAPVC.

- Only patients with isolated supracardiac TAPVC with a discernible ascending vertical vein were included.
- All patients with anomalous pulmonary venous drainage to the right SVC, coronary sinus or right atrium (RA), those with infracardiac and mixed TAPVC or associated complex congenital cardiac malformations and heterotaxy syndromes were excluded.
- All patients with obstructive TPAVC underwent adjustable vertical vein ligation.
- Patients with non-obstructive TAPVC with moderate to severe pulmonary arterial hypertension (PAH) and hemodynamic decompensation, requiring inotropes and ventilator support in the preoperative period underwent adjustable vertical vein ligation (Figure 2).
- In patients with non-obstructive TAPVC, without preoperative PAH or mild PAH, snaring of vertical vein was done after coming...
A 19-year (January 2000 to December 2019) retrospective review of medical records of 99 consecutive patients (62 males) undergoing rechanneling of supracardiac TAPVC with or without vertical vein ligation and adjustable vertical vein ligation at AIIMS, New Delhi operated by a single surgeon (corresponding author) were studied. Inpatient and outpatient records were analysed to obtain demographic, operative, and perioperative features, follow-up echocardiographic computed tomographic (CT) angiographic studies, cardiac catheterization and clinical data.

Operative mortality was defined as patient death either prior to hospital discharge or within thirty days of the date of surgery according to the Congenital Heart Surgery Outcome measures endorsed by the Society of Thoracic Surgeons46.

**Definitions**

**Criteria for pulmonary venous obstruction and indications of cardiac catheterization**

Echocardiography was our principal diagnostic modality, and 60 of the 99 patients proceeded to operation with only echocardiographic data. The indications of preoperative cardiac catheterization were (1) anatomy that was unresolved by echocardiography, (2) characterization of the pulmonary venous obstruction, or (3) exclusion of major associated cardiac anomalies that required delineation or intervention.

Preoperative pulmonary venous drainage was considered obstructive if there was echocardiographic or angiographic data that indicated: (1) a significant gradient between the pulmonary veins and their point of drainage (flow acceleration >2 m/s by echocardiography or pressure gradient >4 mm Hg), (2) monophasic and continuous Doppler flow pattern in the individual pulmonary veins, the pulmonary venous confluence or the vertical vein, or (3) angiographically evident localized reduction in a single pulmonary vein diameter of 50% or more.

Pulmonary hypertension was defined as a right-to-left ventricular systolic pressure ratio of 0.6. According to SPAP, as measured by cardiac

![Diagram of inclusion and exclusion criteria](image)

**Figure 2**: Schematic diagram showing the inclusion and exclusion criteria for selection of patients undergoing rechanneling of supracardiac totally anomalous venous connection (TAPVC) with or without vertical vein ligation and adjustable vertical vein ligation. PAH= pulmonary artery hypertension, SPAP= systolic pulmonary artery pressure, MLAP= mean left atrial pressure.

off cardiopulmonary bypass and systolic PAP (SPAP) and mean LAP (MLAP) were measured. If SPAP ≥31 mmHg, MLAP ≥18 mmHg, and peak systolic right-to-left ventricular pressure ratio ≥1.0, adjustable vertical vein ligation was performed. Otherwise vertical vein was directly ligated and those patients were excluded from the study.
catheterization or on echocardiography on the basis of the velocity of tricuspid regurgitation, pulmonary hypertension was graded as follows: none (SPAP <18 mm Hg), mild (SPAP 19-30 mm Hg), moderate (SPAP 31-50 mm Hg), or severe (SPAP >50 mm Hg.) Isolated or simple TAPVC was diagnosed if the patient had TAPVC in association with a secundum atrial septal defect (ASD), a patent ductus arteriosus, or both. An operation was classified as an emergency if the patient was taken to the operating room within the first 24 hours after arrival at the hospital for hemodynamic or ventilatory compromise.

Low cardiac output syndrome in repaired TAPVC was diagnosed if the patient required inotropic support (dopamine at 4-10 µg/[kg/min]), dobutamine at 5-10 µg/[kg/min], epinephrine at 0.01-0.1 µg/[kg/min], milrinone at 50 µg/kg intravenous bolus followed by 0.375-0.75 µg/[kg/min]), either isolated or in combination in the operating room or in the intensive care unit, to maintain stable hemodynamics in the absence of residual structural lesions and mechanical external compression after correction of all electrolytes or blood gas abnormalities and after adjustment of the preload to its optimal value. Low-output syndrome was also diagnosed if there was an increasing requirement of the previously mentioned inotropes along with afterload reduction with sodium nitroprusside. Patients who received less than 4 µg/kg/min dopamine to increase renal perfusion were not considered to have low output syndrome.

Invasive monitoring to measure cardiac output directly (thermodilution catheter, PA pressure line, and thermistors) is cumbersome and hazardous in children and generally avoided in our setup, except in complex cases. We generally limit intracardiac monitoring to right atrial, LA, and PA pressure lines. Accordingly, under the definition of low output syndrome after repaired TAPVC, an integration of relevant clinical, laboratory, and bedside echocardiographic criteria was used. The criteria for diagnosis were as follows: cold extremities, absent pedal pulses, decreased toe temperature, reduced systolic pressure, impaired renal function and oliguria (<1.0 mL/[kg/hr]), metabolic acidosis, increased serum lactate levels (>2 mmol/L for >2 hours), low mixed venous oxygen saturation (<50%), and blunted sensorium in the absence of residual anatomic or pulmonary venous obstruction.

Postoperative studies

All survivors (n=93) were examined and studied between January 2019 and December 2019, which was the closing interval of the study. Postoperative evaluation consisted of three monthly clinical examination, electrocardiogram, chest radiograph, echocardiography and computerized tomographic angiography. The functional class at follow-up was noted. A minimum of 12 months follow-up was mandatory for echocardiographic and angiographic evaluation. For life table analysis, complete follow-up from the day of operation till the last follow-up was taken into consideration. Computed-tomographic angiography was performed on a third generation, 384 (2 x 192) slice, dual source CT scanner and multiplanar reformatted images and volume rendered images (SOMATOM FORCE, Siemens, Germany) were reconstructed and analysed.

Techniques of Computerised-tomographic Angiography

All scans were performed on a third generation, 384 (2 x 192) slice, dual source CT scanner (SOMATOM FORCE, Siemens, Germany). It has a rotation time of up to 0.25 seconds with a temporal resolution of up to 66 milliseconds and spatial resolution of 0.24 mm.

No form of heart rate control was required. Retrospective ECG-gated CT angiography examination was performed after injection of non-ionic iodinated contrast (1.0 to 1.5 mL/Kg body weight) was administered via peripheral intravenous line at flow rates varying from 1.0 – 4.5 mL/s followed by a saline chaser injected at the same flow rate. A prospectively gated flash mode acquisition at a pitch of 3.2 (table feed 737 mm s⁻¹), and using a tube voltage of 80 kVp with automated tube current modulation was also performed. A “manual” bolus tracking method was used with acquisition manually triggered when optimal contrast opacification within the pulmonary vessels was perceived on the monitoring sequence. Automated tube voltage selection and automated tube current modulation based on body habitus (CARE kV and CARE Dose4D, Siemens Healthcare) were enabled.

Slices were reconstructed of 0.6-mm section thickness and increment of 0.4 mm, using a medium sharp kernel (Bv40), with a model based iterative reconstruction strength level 3 (ADMIRe; Siemens Healthcare). Multiplanar reformatted images and volume rendered images were reconstructed and analysed.

Operative and Postoperative Management

The surgical techniques, intra- and postoperative management protocols were uniform throughout the study period. No patients underwent deep hypothermic circulatory arrest in the study group. Forty-nine (49.4%) patients required emergency operation for hemodynamic or respiratory compromise.

All patients underwent rechanneling of TAPVC with left atrial augmentation, atrial septal fenestration, and adjustable vertical vein ligation under moderately hypothermic CPB and St. Thomas (II) based (4:1) cold blood cardioplegia. Both continuous and modified ultrafiltration was used in all patients to reduce total body water and to remove inflammatory mediators from the circulation. The technical details of the surgical steps including adjustable vertical vein ligation have been enumerated in our previous two publications as well as in the video presentation (Figures 3A-3D, Video presentation) [14-15] We proceeded to keep the vertical vein unligated if the PA pressure remained elevated (systemic or suprasystemic) with a peak systolic right-to-left ventricular pressure ratio ≥ 1.0 after coming off CPB and administration of protamine on snaring the vertical vein. Left atrial and PA pressures were monitored continuously during the operation. After the operation patients were sedated and paralyzed during the first 24 to 48 hours. Pulmonary arterial hypertension was treated with hyperventilation, sedation, phenoxybenzamine, sildenafil citrate and inhaled nitric oxide (10-15 ppm) in varying combination.

After weaning from bypass, upon snaring the vertical vein, the MLAP increased between 18 and 21 mmHg accompanied by an acute increase in systolic PAP to greater than 50 mmHg in 82 (82.8%) patients. Loosening of the vertical vein resulted in a decrease of pressure to a ratio of 0.6 and decrease of the MLAP to 11 mmHg with a range between 10-11 mmHg in all patients. This was associated with a significant increase in mean arterial blood pressure and stable hemodynamics. The PA pressure in the remaining 17 patients was between 31-50 mmHg. The vertical vein was left open on an adjustable vertical vein ligature in all patients.

The sternum was left open in 11 (11.1%) patients. Delayed sternal closure was done 24 to 48 hours after hemodynamic stability was achieved.

Median duration of inotrope requirement was 8 days (range 6-19 days). Median duration of ventilation was 4 days (range 1-20 days). Thirty-nine (39.3%) patients received total parenteral nutrition commencing after 48 hours of ventilation. After the operation, patients were weaned from digoxin, diuretics, phenoxybenzamine and angiotensin-converting enzyme-inhibitors at varying intervals.

The mean CPB time was 56.5±11.9 minutes (range 35-120 minutes), and the mean aortic cross-clamp time was 26.2±8.3 minutes (range 17-70 minutes). Left ventricular assist devices and extracorporeal membrane oxygenation (ECMO) were not used for any patient in this study group.
Figure 3A-3D: Surgical photograph of an adjustable vertical vein ligature (L) showing the sick suture doubly looped around the vertical vein (V) and passed through a polytetrafluoroethylene felt (F). Subsequently, the sutures are passed through the second left intercostal, skin and another polytetrafluoroethylene felt (F) in a straight lie. Two clips (C) are applied individually over each silk suture at the exit point under the subcutaneous tissue. An additional clip is applied over the polytetrafluoroethylene felt which act as a marker. C= Ligaclips, F= polytetrafluoroethylene felt, L= vertical vein ligature, V= vertical vein

Statistical Analysis

Statistical analysis was performed using STATA 14.0 Software (College Station, Texas, USA). Interval related data were expressed as mean ± standard deviation (SD) or median (minimum-maximum) and categorical variables were expressed as percentages. The survival probability with 95% confidence intervals was reported at various time intervals with the Kaplan-Meier technique (Figure 4). The p value of <0.05 was considered as statistically significant.

Figure 4: Actuarial survival curve of patients (n=99) undergoing adjustable vertical vein ligation and left atrial augmentation for obstructive supracardiac totally anomalous pulmonary venous connection.
Results

Original cohort

Patients' age at operation ranged from 1 day to 25 years (mean 24.8±55.3 months, median 4 months). Thirty-five (35.2%) patients were younger than 1 month, 10 (10.1%) were between 6 and 12 months, and 24 (24.2%) were between 12 months and 25 years. Body weight ranged from 1.8 kg to 47 kg (mean 6.3±6.2 kg, median 4 kg). Most patients in this study group were small for age and 60% weighed less than 50th percentile of predicted weight for Indian neonates and infants.

Two-dimensional colour Doppler echocardiography revealed obstructive TAPVC in 49 (49.4%) patients. The level of obstruction was at the insertion site of the vertical vein into the brachiocephalic vein in 25 patients, at the level of left bronchus and left pulmonary artery in 15 patients, and due to restrictive atrial septal defect (ASD) in 9 patients. (Definitions- Electronic)

Ten (10.1%) patients underwent preoperative balloon dilatation of the vertical vein and 9 (9.1%) patients underwent balloon atrial septostomy. The ductus arteriosus was patent in 64 (64.6%) patients. Nineteen (19.2%) patients required preoperative mechanical ventilation, and 32 (32.3%) patients required inotropic support for hemodynamic instability. Their demographic and clinical profiles are depicted in tables 1 and 2.

Cardiac catheterization and angiography were performed in the initial 19 (19.2%) patients in whom an accurate anatomical pattern was unresolved by echocardiography and in 20 (20.2%) patients to evaluate the degree of pulmonary hypertension. Since 2009, computed-tomographic (CT) angiography is used as a routine to delineate the anatomic details in doubtful cases to establish the diagnosis and has almost replaced angiocardiography.

Short-term outcomes

There were 5 (5.0%) operative deaths due to persistent PAH and low cardiac output syndrome (LCOS) (n=2), refractory ventricular arrhythmias (n=1), massive pulmonary haemorrhage (n=1), and sepsis (n=1) during the early period of our study (2002-2010). All survivors exhibited an entirely satisfactory primary repair (echocardiographically, showing a large unrestricted anastomosis, as large as or larger than mitral valve orifice area, with no gradient between the pulmonary venous confluence and LA and non-turbulent biphasic pulmonary venous flow at ≤1.2 m/sec. Ten (10.1%) patients demonstrated subtle echocardiographic changes with a large, unrestricted anastomosis with no gradient between the common pulmonary venous chamber and LA and biphasic or mainly biphasic pulmonary venous flow with a velocity of 1.2 to 1.6 m/sec.

Despite having an unligated vertical vein, 13 (13.1%) patients with obstructive TAPVC with PAH undergoing emergency surgical intervention for hemodynamic or respiratory compromise exhibited suprasystolic PAP and unstable postoperative hemodynamics. Among them, the sternum had to be left open in 11 patients. Delayed sternal closure was done in 24-48 hours after achieving stable hemodynamics. These patients were mechanically ventilated with an inspired oxygen fraction of 0.8 and 20 ppm nitric oxide and were administered pulmonary vasodilators (sodium nitroprusside 0.5µg/Kg/min, phenoxybenzamine 0.5 µg/kg at 8-hour intervals) for 15-20 days. Intraoperative transesophageal echocardiography at the time of sternal closure continued to demonstrate shunting from the common pulmonary venous chamber (CPVC) to the brachiocephalic vein and attempted snaring of the vertical vein resulted in rise of PAP to systemic and suprasystolic levels in all of them. Despite all attempts, 2(2%) patients died of pulmonary hypertensive episodes.

Serial echocardiography during these episodes demonstrated shunting from the CPVC to the brachiocephalic and right-to-left shunting across the fenestrated atrial septal patch, clearly documenting their roles as a temporary vent (Figures 5A, 5B, 6A-6D).

Postoperatively, all patients underwent daily echocardiographic assessment of biventricular function and flow through the vertical vein. Serial postoperative echocardiograms showed unobstructed pulmonary venous return into LA with a distented vertical vein (Figures 5A, 5B).

Figures 5A, 5B: Postoperative transthoracic two-dimensional echocardiogram: 2A: Day 1 postoperative with color flow patterns of venous flow in the unligated vertical vein showing shunting from the left atrium to the brachiocephalic vein via the patent vertical vein; 2B: Demonstration of change of the shunting patterns on the 10th postoperative day on the same patient from the brachiocephalic vein to the left atrium via the vertical vein.

Although all survivors were extubated within 24 to 96 hours with stable hemodynamics, 35 (35.35%) patients continued to have tachypnoea and biventricular failure during the first two weeks. Postoperatively, serial echocardiography demonstrated reversal of flow from the brachiocephalic vein to LA in all patients. The vertical vein was interrupted in all patients using the adjustable vertical vein ligature when there was phasic reversal of flow from the brachiocephalic vein to the LA on color Doppler (Figures 5A, 5B, 6A-6D).
Figures 6A-6D: Postoperative transthoracic two-dimensional echocardiogram showing the newly constructed interatrial septal patch with left atrial augmentation and atrial septal fenestration (arrow). Doppler flow shows there is complete interruption of blood flow through the vertical vein after adjustable vertical vein ligation. (AoV= Aortic valve, CC= Common pulmonary venous chamber, LA Aug= Augmented left atrium, PA= Pulmonary artery, RA= Right atrium, VV= Vertical vein).

At this point, we decided to tighten the adjustable ligature in increments. Tightening was achieved by placing additional clips outside between the polytetrafluoroethylene pledget and the previous clip, while monitoring the LA and PA pressure and arterial blood gases. Echocardiographic assessment was performed to assess the ventricular function and the degree of tightening was assessed by reduction of left-to-right shunt through the vertical vein. All ligatures were tightened gradually over a period of 24 to 96 hours, maintaining stable hemodynamics with PA pressure at subsystemic levels and normal blood gases.

**Long-term outcomes**

There were 2 (2.0%) late deaths 80 and 94 months after surgery due to ventricular arrhythmia (n=1) and pulmonary sepsis (n=1) respectively. Follow-up was 100% complete (1-228 months) and yielded 1695.7 patient-years of data. At a mean follow-up of 218.8 (SE± 209.9) months, the actuarial survival was 93.7 (SE±0.02%; 95% CI: 86.5, 97.1; Figure 4). All survivors were in New York Heart Association I or II at their last follow-up. There were no reoperations during this period.

At a mean follow-up of 218.8 (±SE, 209.9) months, serial 2-dimensional echocardiography did not reveal any evidence of pulmonary venous obstruction, or residual flow through the ligated vertical vein (Figures 5A, 5B, 6A-6D).

Angiographically, none of the survivors demonstrated any flow through the vertical vein and none had evidence of restenosis of the common pulmonary venous chamber-LA anastomosis. There was marked increase in the volume of the left atrium and there was no distortion of the left superior pulmonary vein and left brachiocephalic vein on any patient (Figures 7A-7D, 8A-8D, 9A-9C, 10A-10D)
Figures 7A-7D: Axial images and three-dimensional volume rendered computed tomographic image in the oblique coronal projections (A and B; preoperative) and (C and D; postoperative) showing marked increase in the volume of the left atrium from 7.3 ml (preoperative) to 9.3 ml (postoperative) due to left atrial augmentation and changes in the loading condition of the left heart following common pulmonary venous chamber to left atrial anastomosis. (LA=left atrium)

Figures 8A-8D: Postoperative preligation, three-dimensional volume rendered coronal computerized tomographic image- A) anterior, B) posterior view showing unligated vertical vein and the wide unrestricted anastomosis between the common pulmonary venous chamber and left atrial appendage, C) anterolateral, & D) posterior projection showing the cranial part of the ligated vertical vein and all pulmonary veins draining into the common pulmonary venous chamber which has been anastomosed to the left atrial appendage (CC= Common pulmonary venous chamber, DTA= Descending thoracic aorta, LAA= Left atrial appendage, LSPV= Left superior pulmonary vein, RIPV= Right inferior pulmonary vein, RSVC= Right superior vena cava, RSPV= Right superior pulmonary vein, SVC= Superior vena cava).
Figures 9A-9C: Postoperative postligation, three-dimensional volume rendered computed tomographic images in the coronal (A), left anterior oblique projection (B) and the postero-anterior projection (C) showing the cranial part of the ligated vertical vein and the wide unrestricted anastomosis between the common pulmonary venous chamber and left atrial appendage. All pulmonary veins are draining into the common pulmonary venous chamber which has been anastomosed to the left atrial appendage (CC= Common pulmonary venous chamber, DTA= Descending thoracic aorta, LAA= Left atrial appendage, SVC= Superior vena cava).

Figures 10A-10D: Postoperative postligation, axial image (A), coronal image, three-dimensional volume rendered computed tomographic image in the postero-anterior projection (C) and antero-posterior projection (D) showing the cranial part of the ligated vertical vein and the wide unrestricted anastomosis between the common pulmonary venous chamber and left atrial appendage. All pulmonary veins (*) are draining into the common pulmonary venous chamber which has been anastomosed to the left atrial appendage. (CC= Common pulmonary venous chamber, LA= Left atrium, LV=left ventricle, LBCV= Left brachiocephalic vein, LSPV= Left superior pulmonary vein, SVC= Superior vena cava; VV= Vertical vein).
Comment

The medical literature is rife with examples of therapies that seemed likely to be beneficial, but were in fact either pointless or harmful. As far as we could establish, there have been limited studies addressing the usefulness of vertical vein interruption at the time of TAPVC repair and the fate of unligated vertical vein following repair of TAPVC. There remain several unresolved queries in the medical and surgical management of TAPVC and there is no fool proof formula in the published literature to decide the optimal surgical management for a given patient.

Our 99-patient cohort is perhaps the largest so far to be evaluated for selective vertical vein patency, adjustable vertical vein ligature, LA augmentation and atrial septal fenestration on the postoperative outcomes of repaired TAPVC The age distribution was heterogenous with 35 (35.2%) patients being ≤ 1 month, and 24 (24.2%) were between 12 months and 25 years. The patient’s socioeconomic profiles and lack of healthcare access led to their referrals for surgery at older ages.

In our earlier publications, we demonstrated that late referral and late presentation lead to the development of severe PAH, malnutrition and ultimately cardiac cachexia. These factors predispose them to pulmonary infection, sepsis, postoperative pulmonary hemorrhage, and unfavourable reactions to bypass related stresses. Despite aggressive perioperative management, we reported worse outcomes in whom conventional strategy of routine ligation of a discernible ascending or descending vertical vein was performed.

One important finding of our investigation was the occurrence of post bypass suprasystemic PA pressure due to late presentation, cardiac cachexia and cardiac pathology.

Our second finding is the occurrence of suprasystemic PA pressure and unstable hemodynamics upon snaring the vertical vein after weaning from bypass. Release of the snare resulted in improved postoperative haemodynamics, early weaning from ventilatory support, and survival. In the absence of anatomic sticture, this undesirable effect can be explained by impaired unloading of the pulmonary venous chamber due to non-compliant left-sided cardiac chambers.

The benefits of an unligated vertical vein over the traditional concept of routine vertical vein ligation at the time of TAPVC repair remains debatable. As yet, there are no universally accepted criterions for selection of patients in whom it is advantageous to maintain the patency of the vertical vein.

A number of studies with small cohort of patients have been published by various investigators with conflicting results (Table 3) [16-23]. Proponents of leaving the vertical vein unligated presume that it helps to tide over postoperative pulmonary hypertensive crisis [13,18,20,21]. Furthermore, some investigators have shown that the flow through vertical vein ceases as time passes and there is no requirement for further intervention [13,17,22]. However, there are few reports of persistent left-to-right shunt through the vertical vein leading to cardiac failure, which required reoperation for ligation of vertical vein [17,18,20].

Advocates of vertical vein ligation even in patients with moderate or severe pulmonary hypertension rely on advanced postoperative intensive care management utilizing nitric oxide therapy, ventilator management strategies and occasionally on ECMO [18,23,24]. Recurrent episodes of pulmonary hypertensive crises, rapid development of pulmonary arterial and venous medial hypertrophy, and a small noncompliant LA and LV have been variously implicated as the causative factors for the LCOS [1-4,14,25-32].

In 2007, we demonstrated that acute vertical vein ligation resulted in elevated LA pressure and impaired LV function suggesting that for a period of time the small, poorly compliant LV of the patient with obstructive TAPVC was unable to maintain adequate cardiac output [14].

The unligated vein group in our previous study demonstrated statistically significant decreased in-hospital mortality (p=0.04), decreased pulmonary hypertensive crises (p<0.001), shortened intensive care and hospital stay (p<0.001) and superior postoperative hemodynamics (p<0.001). Logistic regression analysis demonstrated 3.28 times (95% CI: 108.9-9.99) increased risk of death after vertical vein ligation [14].

Contrary to the reports by Cope and colleagues in which patent vertical vein atrophied, 11 of 23 survivors in our previous investigation had symptoms of a large left-to-right shunt through the unligated vertical vein requiring delayed interruption. The vertical vein was ligated through resternotomy in 4 patients, left anterolateral thoracotomy in 2 cases and adjustable vertical vein ligature in 5 patients [13,14]. Due to non-availability of Amplatzer vascular plug, these six patients underwent surgical ligation of the vertical vein. Although delayed closure of the vertical vein was successful in all cases it was attended by extremely high PAP in six patients and proved a difficult postoperative challenge. These findings were suggestive of a relatively small, non-compliant dysfunctional left-sided chambers or of disease-related or CPB-related pulmonary vasoreactivity [14].

There are varying reports on left heart volume characteristics in obstructive TAPVC from normal findings to the conclusion that left-sided chambers are smaller than normal and the LA lacks both normal compliance and reservoir function [5-11]. Such chamber abnormalities have been attributed to large left-to-right shunt causing reduced atrial filling and decreased LV relaxation secondary to elevated right ventricular diastolic pressure or volume [5-11]. Published literature does not address the percentage of patients with obstructive TAPVC who have structurally smaller left-sided chambers.

In order to allow the left heart to adapt and maintain adequate cardiac output, we have used an oversized Dacron patch for interatrial septation, deviating the same towards the enlarged right atrium and have incorporated part of the vertical vein to achieve structural alignment and augmentation of the LA cavity (Figures 7A-7D) [14]. The concept of enlarging the LA is almost similar to that described by Cooley and colleagues except that in our patients, we achieved it by excising the floor of the fossa ovalis and utilizing an oversized Dacron patch graft to close the ASD and enlarge the LA [33].

Corno and colleagues described the two-patch technique of LA enlargement. Subsequently, several investigators have demonstrated increased incidence of supraventricular arrhythmias due to the use of a transverse right atrial incision and division of the supraventricular crest. We have not used this technique. There is no valid argument for or against the use of Dacron patch or pericardial patch. It may be argued that fenestration in the synthetic patch may be appropriately sized at the primary operation with minimal chances of over or undersizing. There were no residual shunts through the fenestration on any surviving patient on long-term follow-up.

The driving forces behind selection of patients whose vertical vein was kept patent after surgery were to reduce the PAP in the perioperative period, postoperative pulmonary hypertensive crises and mortality (Figure 2). We aimed to achieve this by leaving the vertical vein unligated so that it may function as a temporary venous reservoir for pulmonary venous blood, volume unloading the small non-compliant left-sided cardiac chambers until they are able to grow and adapt to the requisite flow demands. Secondly, the unligated vertical vein could serve as a temporary “pop-off” valve in the event of pulmonary hypertensive crises and thereafter the same may be subjected to a gradual process of occlusion. Finally, despite late presentation and long standing pulmonary hypertension, we have been successful in occluding the patent vertical vein within 72 to 96 hours. (Figures 5A, 5B, 6A-6D).
The etiopathogenesis of postoperative pulmonary hypertensive crises in obstructive TAPVC may be multifactorial [25-32]. Although the pulmonary veins/capillaries do not have any valves, there are variable reactive pulmonary arteriolar changes due to the disease process and any rise in LAP will lead to pulmonary hypertensive crisis. The postoperative course targets management of these issues and occasionally ECMO is necessary while the pulmonary vascular bed recovers and LV compliance improves [14,15,24]. The time course of this adjustment is variable. It is conjectured that there is a feedback loop whereby acute elevation of LAP exacerbates the low cardiac output state caused by a poorly compliant LV [14].

Atrial septal fenestration in patients undergoing rechanneling of TAPVC is controversial. In this study, we performed atrial septal fenestration to permit “spill over” in the setting of postoperative RV dysfunction in patients with both obstructive and non-obstructive supracardiac TAPVC with PAH. During episodes of pulmonary hypertensive crises with limited RV output, and an elevated central venous pressure (CVP), a fenestrated atrial septal patch permitted right-to-left shunting, increasing left ventricular preload and cardiac output, albeit at the expense of some degree of systemic desaturation (Figures 5A, 5B, 6A-6D). It is noteworthy that none of the survivors exhibited any shunting in the late postoperative period and none required any interventional closure of the fenestration. Thus, the unligated vertical vein, in conjunction with a calibrated atrial septal fenestration, resulted in equalization of LAP and CVP during episodes of pulmonary hypertensive crisis, and was the automatic choice to avert a dismal outcome in the perioperative period. Our protocol is to institute mechanical circulatory assistance, if the cardiac output cannot be sustained by the currently available medical and surgical treatment. We have not used ECMO on any of our patients in this study group.

<table>
<thead>
<tr>
<th>Patient related variables</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at operation (months, range, median)</td>
<td>24.8±55.3 months, range 1 day-25 years, median 4 months</td>
</tr>
<tr>
<td>Age distribution</td>
<td></td>
</tr>
<tr>
<td>&lt;1 month</td>
<td>35 (35.35%)</td>
</tr>
<tr>
<td>1-6 months</td>
<td>30 (30.3%)</td>
</tr>
<tr>
<td>6-12 months</td>
<td>10 (10.1%)</td>
</tr>
<tr>
<td>12 months-25 years</td>
<td>24 (24.2%)</td>
</tr>
<tr>
<td>Weight (kg, mean±SD, range, median)</td>
<td>6.3±6.2 kg (range 1.8-47 kg, median 4 kg)</td>
</tr>
<tr>
<td>Anatomies Supracardiac with ascending vertical vein</td>
<td>99 (100%)</td>
</tr>
<tr>
<td>Supracardiac totally anomalous pulmonary venous connection Obstructive Non-obstructive</td>
<td>49 (49.5%), 50 (50.5%)</td>
</tr>
<tr>
<td>Preoperative pulmonary arterial hypertension</td>
<td>99 (100%), 0</td>
</tr>
<tr>
<td>Preoperative inotropes</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>32 (32.3%)</td>
</tr>
<tr>
<td>No</td>
<td>67 (67.7%)</td>
</tr>
<tr>
<td>Preoperative ventilation</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>19 (19.2%)</td>
</tr>
<tr>
<td>No</td>
<td>80 (80.8%)</td>
</tr>
<tr>
<td>Operation timing</td>
<td></td>
</tr>
<tr>
<td>Emergency</td>
<td>49 (49.5%)</td>
</tr>
<tr>
<td>Elective</td>
<td>50 (50.5%)</td>
</tr>
<tr>
<td>Mean cardiopulmonary bypass time (range)</td>
<td>56.5±11.9 (35-120 minutes)</td>
</tr>
<tr>
<td>Mean aortic cross-clamp time (range)</td>
<td>26.2±8.3 (17-70 minutes)</td>
</tr>
<tr>
<td>In-hospital mortality</td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>5 (5.1%)</td>
</tr>
<tr>
<td>Cardiac and pulmonary hypertension related</td>
<td>4 (4.0%)</td>
</tr>
<tr>
<td>Late death</td>
<td>2 (2.0%)</td>
</tr>
<tr>
<td>Pulmonary hypertensive crisis</td>
<td>16 (16.1%), 83 (83.8%)</td>
</tr>
<tr>
<td>Low cardiac output</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>58 (58.6%)</td>
</tr>
<tr>
<td>No</td>
<td>41 (41.4%)</td>
</tr>
<tr>
<td>Duration of inotrope requirement</td>
<td></td>
</tr>
<tr>
<td>Median (range)</td>
<td>8 (6-19 days)</td>
</tr>
<tr>
<td>Duration of ventilation</td>
<td></td>
</tr>
<tr>
<td>Median (range)</td>
<td>4 (1-20 days)</td>
</tr>
</tbody>
</table>

Table 1: Demographic, operative and postoperative details and the study group (n=99)
### Patient related variables

<table>
<thead>
<tr>
<th></th>
<th>Obstructive</th>
<th>Non obstructive</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>49 (49.5%)</td>
<td>50 (50.5%)</td>
</tr>
<tr>
<td>Mean age at operation, months, range</td>
<td>1.94±1.8</td>
<td>22.6±51.8</td>
</tr>
<tr>
<td>Age distribution</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1 month</td>
<td>35 (71.4%)</td>
<td>0</td>
</tr>
<tr>
<td>1-6 months</td>
<td>14 (28.6%)</td>
<td>16 (32%)</td>
</tr>
<tr>
<td>6-12 months</td>
<td>0</td>
<td>10 (20%)</td>
</tr>
<tr>
<td>12 months-25 years</td>
<td>0</td>
<td>24 (48%)</td>
</tr>
<tr>
<td>Anatomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Supracardiac totally anomalous venous connection with ascending vertical vein</td>
<td>49 (100%)</td>
<td>50 (100%)</td>
</tr>
<tr>
<td>Preoperative pulmonary arterial hypertension</td>
<td>49 (100%)</td>
<td>50 (100%)</td>
</tr>
<tr>
<td>Yes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>32 (65.3%)</td>
<td>0</td>
</tr>
<tr>
<td>Preoperative inotropes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>19 (38.7%)</td>
<td>0</td>
</tr>
<tr>
<td>No</td>
<td>30 (61.3%)</td>
<td>50 (100%)</td>
</tr>
<tr>
<td>Preoperative ventilation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>45 (91.8%)</td>
<td>4 (8%)</td>
</tr>
<tr>
<td>No</td>
<td>4 (8.2%)</td>
<td>46 (92%)</td>
</tr>
<tr>
<td>Operation timing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Emergency</td>
<td>53.8±10.6 (35-100 min)</td>
<td>58.7±12.9 (41-120 min)</td>
</tr>
<tr>
<td>Elective</td>
<td>58.7±12.9 (41-120 min)</td>
<td>53.8±10.6 (35-100 min)</td>
</tr>
<tr>
<td>Mean aortic cross-clamp time (range)</td>
<td>25.7±6.8 (17-50 min)</td>
<td>28.2±9.3 (37-70 min)</td>
</tr>
<tr>
<td>In-hospital mortality</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac and pulmonary hypertension related</td>
<td>2 (4%)</td>
<td>2 (4%)</td>
</tr>
<tr>
<td>Late death</td>
<td>1 (2%)</td>
<td>1 (2%)</td>
</tr>
<tr>
<td>Pulmonary hypertensive crisis</td>
<td>10 (20.4%)</td>
<td>6 (12%)</td>
</tr>
<tr>
<td>Yes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Low cardiac output</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>40 (81.6%)</td>
<td>18 (36%)</td>
</tr>
<tr>
<td>Duration of inotrope requirement</td>
<td>10 (6-10 days)</td>
<td>6 (1-6 days)</td>
</tr>
<tr>
<td>Median (range)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Duration of ventilation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median (range)</td>
<td>4 (1-20 days)</td>
<td>3 (1-5 days)</td>
</tr>
</tbody>
</table>

### Table 2: Demographic, operative and postoperative details of the study group (Obstructive vs. non-obstructive: n=99)

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Author(s)</th>
<th>Year of study</th>
<th>Diagnosis</th>
<th>Number of patients</th>
<th>Age/Weight</th>
<th>Unligated vertical vein (Number)</th>
<th>Follow-up</th>
<th>Results</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Cope J et al&lt;sup&gt;1,13&lt;/sup&gt;</td>
<td>1974-1995</td>
<td>Supracardiac TAPVC obstructive (n=4)</td>
<td>4</td>
<td>1 day-5 years</td>
<td>4</td>
<td>1 day-106 months</td>
<td>All had patent vertical vein on follow-up</td>
<td>All survived</td>
</tr>
<tr>
<td>2.</td>
<td>Zhao K et al&lt;sup&gt;17&lt;/sup&gt;</td>
<td>1982-2008</td>
<td>Supracardiac (n=77)</td>
<td>20 unligated</td>
<td>8.5±7.0 years 27 days-44 years</td>
<td>20</td>
<td>1 day-5 years</td>
<td>Spontaneous closure (n=8) Remained patient (n=12)</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>Kelle AM, et al&lt;sup&gt;12,3&lt;/sup&gt;</td>
<td>1990-2008</td>
<td>Supracardiac (n=39) obstructive (n=22)</td>
<td>39 ligated</td>
<td>0-1.7 years (range 14.6 days (median)</td>
<td>39 (used nitric oxide in addition)</td>
<td>4.21±5.2 years (1 month-18.6 years)</td>
<td>Vertical vein ligated in all patients + nitric</td>
<td></td>
</tr>
</tbody>
</table>

Auctores Publishing – Volume 3(8):075 www.auctoresonline.org
ISSN:2641-0419

Page 12 of 16
<table>
<thead>
<tr>
<th>Study</th>
<th>Reference</th>
<th>Period</th>
<th>Condition</th>
<th>Number of Patients</th>
<th>Vertical Vein Ligature</th>
<th>Duration of Intubation</th>
<th>Weight (kg)</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>4.</td>
<td>Caspi J et al&lt;sup&gt;18&lt;/sup&gt;</td>
<td>1993-2000</td>
<td>Supracardiac TAPVC obstructive (n=9)</td>
<td>3</td>
<td>3.5 kg</td>
<td>3 days</td>
<td>3</td>
<td>(8-71 months) 38±6 months</td>
<td>Closed in ICU in all patients, tightened on snare</td>
</tr>
<tr>
<td>5.</td>
<td>Kumar S, et al&lt;sup&gt;19&lt;/sup&gt;</td>
<td>1995-1999</td>
<td>Supracardiac TAPVC obstruction</td>
<td>10</td>
<td>5-80 days</td>
<td>Weight 3-4.5 kg</td>
<td>3</td>
<td>6 months-2 years</td>
<td>All patient at follow-up No spontaneous closure</td>
</tr>
<tr>
<td>6.</td>
<td>Saritas B, et al&lt;sup&gt;21&lt;/sup&gt;</td>
<td>1996-2010</td>
<td>Supracardiac TAPVC obstruction</td>
<td>14</td>
<td>3.8±2.1 months</td>
<td>Weight 4.4±1.1 kg</td>
<td>14</td>
<td>48±36 months</td>
<td>Spontaneous closure (n=1) Surgical closure (n=2)</td>
</tr>
<tr>
<td>7.</td>
<td>Shah MJ et al&lt;sup&gt;20&lt;/sup&gt;</td>
<td>2000</td>
<td>Supracardiac TAPVC obstructive (n=2)</td>
<td>2</td>
<td>2.5 months/3.5 kg 2 months/4.0 kg</td>
<td>2</td>
<td>5 months and 7 months</td>
<td>Both survived Vertical vein ligated in both patients</td>
<td></td>
</tr>
<tr>
<td>8.</td>
<td>Chowdhury UK et al&lt;sup&gt;14&lt;/sup&gt;</td>
<td>1997-2006</td>
<td>Supracardiac TAPVC obstructive</td>
<td>30</td>
<td>1 day-8 months (range) Mean 1.49±1.63 months Median 1 month Weight: 3.75±1.08 kg</td>
<td>30</td>
<td>1 month-104 months (range) Mean 33.34±29.88 months Median 30 months</td>
<td>Vertical vein was ligated in all patients- approaches resternotomy, left anterolateral thoracotomy, adjustable vertical vein ligature</td>
<td></td>
</tr>
<tr>
<td>9.</td>
<td>Chowdhury UK et al&lt;sup&gt;15&lt;/sup&gt;</td>
<td>2007</td>
<td>Supracardiac obstructive</td>
<td>5</td>
<td>2, 4, 3, 4 and 3 months Weight: 3.2, 4.0, 3.6, 4.2 and 3.4 kg</td>
<td>5</td>
<td>2-107 months Mean: 55.4±45.5 months</td>
<td>Adjustable vertical vein ligature. Gradually tightened over 24-96 hours</td>
<td></td>
</tr>
</tbody>
</table>

TAPVC=Totally anomalous pulmonary venous connection

**Table 3:** Summary of the published investigations documenting the fate of a patent vertical vein in the setting of obstructive supracardiac totally anomalous pulmonary venous connection (TAPVC)

Based on the literature and reasoning cited above, we decided to device a method to avail the benefits of an unligated vertical vein in patients with obstructive TAPVC with post-bypass systemic or suprasystemic PAH, at the same time avoid the nuances of residual left-to-right shunt in the late follow-up when the period of acute crisis is over. The technique and the initial observations following adjustable ligature of the vertical vein was published in 2007 [15].

Thus, the advantages of adjustable vertical vein ligature were:

- The facility to tighten the ligature under optimal physiologic conditions, concomitant with the disappearance of disease or bypass-related pulmonary reactivity.
- The ability to gradually increase the afterload to the ventricle as tolerated by the patient without causing haemodynamic instability under optimal physiologic conditions.
- The capability to occlude the vertical vein in the event of a significant left-to-right shunt and bi-ventricular failure without re-

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opening the sternum or employing another thoracotomy or transcatheater vein closure.

The drawback of this technique may be the inability to loosen the ligature in the event of hemodynamic decompensation. All ligatures in this cohort were tightened gradually in increments over a period of 24-96 hours maintaining stable hemodynamics.

A persistent left-to-right shunt with bi-ventricular failure through an unligated vertical vein does not necessarily relegate a patient to a second-stage operation and does not warrant modification of our selection criteria for the unligated vertical vein. They may be candidates for adjustable vertical vein ligature or transcatheater vertical vein closure [15-35].

An initial concern about the technique was the possibility of iatrogenic distortion of the left superior pulmonary vein and left brachiocephalic vein. In order to address these concerns, we have threaded the loop ligature through a polytetrafluoroethylene felt and secured it to adventitia of the vertical vein, thus preventing its displacement, and brought the arms of the silk suture through the second left intercostal space, away from the sternotomy incision, perpendicular to the vertical vein, ensuring a vertical straight lie, and avoiding distortion or occlusion of the left upper pulmonary and brachiocephalic veins (Figures 3A–3D). Postoperatively, we performed computed-tomographic angiography on all survivors at follow-up and discovered no untoward findings (Figures 8A–8D, 9A–9C, 10A–10D).

Vv-ligature(Video).mp4

Figure Legends of Video Presentation

A video presentation of the surgical techniques of adjustable vertical vein ligation and left atrial augmentation in obstructive supracardiac totally anomalous pulmonary venous connection (UKC’s modification).

1. Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein. The pericardium was opened in the midline in between stay sutures using scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation.

2. The operation was performed with moderately hypothermic cardiopulmonary bypass through angled venous cannula into the superior SVC, straight venous cannula into the IVC, and aortic cannulation. St. Thomas-II based cold hyperkalemic blood cardioplegia (1:4) and topical hypothermia were used for myocardial preservation.

3. The persistent ductus arteriosus was ligated using No.2 ductus silk suture pulling down the superior surface of the pulmonary artery at the commencement of cardiopulmonary bypass as described by Dwight McGoan. The pump flow was temporarily lowered at the time of ligation of the ductus arteriosus.

4. During the cooling phase, the vertical vein was dissected extrapericardially and looped taking care not to snare until the rechanneling was completed. Following aortic cross-clamp and administration of cardioplegia, the right pleural cavity was widely opened. The apex of the heart was lifted cephalad and to the right and dislocated into the right pleural cavity to facilitate surgical exposure.

5. A stay suture was being placed on the under surface of the left pulmonary artery and put on traction for improvement of exposure of the common pulmonary venous chamber. The tip of the left atrial appendage was tied using 2-0 silk sutures and placed on traction.

6. Long transverse incisions were made on the anterior surface of the common pulmonary venous chamber along with the length of the common pulmonary vein and the posterior surface of the left atrial appendage and body of the left atrium in between stay sutures of 6-0 polypropylene (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, USA).

7. The common pulmonary venous chamber and the left atrium were anastomosed using 5-0 polypropylene suture. Precautions were taken to create a large anastomoses (2.5–3.0 cm) or as large as the calculated mitral valve orifice area.

8. The heart was relocated back into the pericardial cavity. The right atrium was opened in between stay sutures 1 cm posterior and parallel to the right atrioventricular groove. The remnant of the septal tissue within the fossa ovalis was excised; the size of the atrial septal defect was measured.

9. An oversized Dacron polyester patch little larger than the size of the atrial septal defect was sutured to close the atrial septal defect in such a way that the right side of the patch was deviated to the body of the right atrium away from the margin of the atrial septal defect. This manoeuvre enhances the capacity of the left atrium.

10. A 2mm calibrated atrial septal fenestration was being performed for decompression of the right-sided chambers in the event of pulmonary hypertensive crisis.

11. The right atrium was closed in two layers using 5-0 polypropylene. The aortic cross-clamp was being released, thus restoring blood flow to the myocardium.

12. A right angle forceps was passed around the vertical vein between its junctions with the left upper pulmonary vein and brachiocephalic vein and is looped using No.4 SUTUPAK suture (Ivory Braided Silk, SW 218, Johnson and Johnson Inc, Somerville, NJ).

13. The right angled forceps was passed again in a similar fashion, and the suture was grasped by the tip of the forceps so that the vertical vein was doubly looped.

14. Both the ends of the thread were next passed through a 0.5 cm x 0.5 cm low porosity No.1 Bard polytetrafluoroethylene pledget (Impra Inc, A Subsidiary of CR Bard, Tempe, Ariz, USA) which was anchored to the adventitia of the vertical vein with interrupted 6-0 polypropylene sutures to prevent subsequent distortion or occlusion of the left upper pulmonary vein and brachiocephalic vein.

15. Both arms of the silk suture were then brought out through the second left intercostal space away from the sternotomy incision perpendicular to the vertical vein, ensuring a vertical, non-redundant straight lie without distorting the vertical vein.

16. The two ends of the sutures were next passed through a 2 x 2 cm polytetrafluoroethylene pledget and clipped together using a big ligaclip (LT 400, Ethicon, Endosurgery Inc, Cincinnati, Ohio). The chest was primarily closed in layers.

17. The percutaneously adjustable device to ligate the vertical vein allows gradual tightening or loosening of the ligature under optimal physiologic conditions, without reoperating the sternum, or having to resort to another thoracotomy once the reactive components of the pulmonary hypertension disappear.

Study limitations

This study has several limitations. In as much as this is a single-surgeon, single-center study, the results may not necessarily be applicable to all centers. Given its retrospective design, there may have been unmeasured confounding factors that influence the observed outcomes. The study institution likely has a referral bias for high-risk patients with complex congenital heart diseases, prematurity, late presentation, and referral from outside the geographical area.
Conclusions

We conclude that in obstructive and non-obstructive supracardiac TAPVC, postoperative pulmonary hypertension is common, and augmenting left atrial compliance by atroplasty is perhaps useful. By leaving a pop-off, either an atrial septal fenestration, and/or the vertical vein in the immediate postoperative period, can be helpful to maintain cardiac output when there is an expected pulmonary hypertensive crisis. One or both of these pop-offs will at some point become redundant, with shunting only left-to-right, which slowly but surely, will lead to volume overload, and they will need to be addressed. In a setting where transcatheter solutions (plugging a vertical vein or ASD fenestration with a device) are not available or prohibitively expensive, percutaneous adjustable vertical vein ligature is a useful adjunct.

It allows gradual tightening of the ligature under optimal physiologic conditions without reoperation and without causing any distortion of the left superior pulmonary vein and left brachiocephalic vein. Knowledge of this approach should contribute to the armamentarium of cardiac surgeon faced with obstructive TAPVC with pulmonary arterial hypertension.

Compliance with ethical standards

Statement of human rights/ethical approval

The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki declaration of 1975, as revised in 2008 and has been approved by the Institutional Research Committee.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of the article.

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References


