



Operative, Echocardiographic and Angiocardiographic Details of the Vertical Vein in Repaired Supracardiac Obstructive Totally Anomalous Pulmonary Venous Connection: Pre- and Post-Adjustable Vertical Vein Ligature

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Abstract

A 2-years-old child with obstructive supracardiac Totally Anomalous Pulmonary Venous Connection (TAPVC) underwent rechanneling of supracardiac TAPVC with an unligated vertical vein for post bypass supra-systemic pulmonary artery pressure. We describe here a new device, permitting adjustable ligation of the vertical vein which permitted us to avoid multiple reoperations. Computed tomographic angiograms during follow-up revealed absence of flow through the vertical vein and ruled out distortion of the left superior pulmonary and left brachiocephalic veins.

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Introduction

Postoperative pulmonary hypertensive crises, right-sided cardiac failure and low cardiac output syndrome remain the predominant causes of death in patients undergoing rechanneling of supracardiac obstructive Totally Anomalous Pulmonary Venous Connection (TAPVC) [1-4]. Whether an adjustable vertical vein ligature with concomitant rechanneling of supracardiac obstructive TAPVC is advantageous over the traditional concept of routine vertical vein ligation is a subject of debate [4-16]. Since all investigators and surgeons have not accepted these findings or utilized these techniques of adjustable vertical vein ligature, we present herein the technical details, echocardiographic and angiocardiographic appearance of the patent and ligated vertical vein in a surviving patient undergoing rechanneling of obstructive supracardiac TAPVC.

Case Presentation

A 2-years-old boy, weighing 5.0 kg was referred to our cardiac clinic for evaluation. He was born the third child of a non-consanguineous marriage and the mother underwent a full-term normal pregnancy and delivery. The child had feeding difficulty with repeated episodes of chest infection. Clinically, he had cardiomegaly, left parasternal heave and loud pulmonic component of second heart sound. The non-invasive oxygen saturation at rest was 80% as determined by pulse oxymetry. Chest radiography revealed cardiomegaly with a cardiothoracic ratio of 0.7, mainly involving the right ventricle and equalization of pulmonary venous congestion. Cross-sectional echocardiography established the diagnosis of obstructive supracardiac TAPVC with the pulmonary veins draining via vertical vein into brachiocephalic vein. The child was referred for rechanneling of TAPVC.

Totally anomalous pulmonary veins draining to the right superior caval vein via the vertical vein and the left brachiocephalic vein was diagnosed during surgery. A wide unrestricted anastomosis between the common pulmonary venous chamber and left atrial appendage was performed via posterior approach and the left atrium was augmented via right atrium using a redundant Dacron patch. The Dacron patch was fenestrated. After weaning from bypass, upon snaring the vertical vein, the median left atrial pressure increased to 22 mmHg, and ranged from 18 to 24 mmHg accompanied by an acute increase in pulmonary artery pressure to suprasystemic levels. Loosening

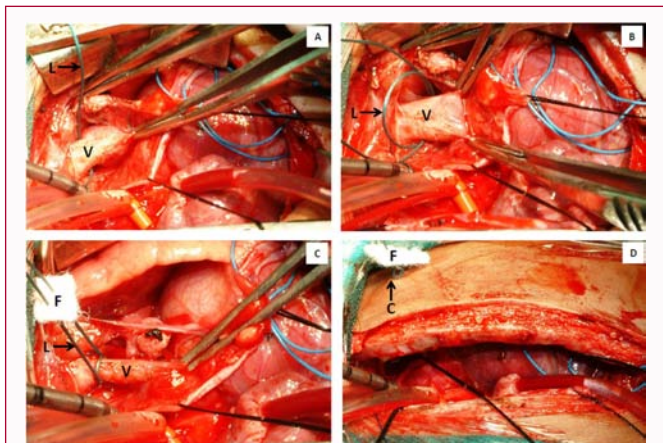


Figure 1A-1D: Surgical photograph of an adjustable vertical vein ligature (L) showing the silk suture doubly looped around the vertical vein (V) and passed through a polytetrafluoroethylene felt (F). Subsequently, the sutures are passed through the second left intercostal, skin and another polytetrafluoroethylene felt (F) in a straight line. 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.

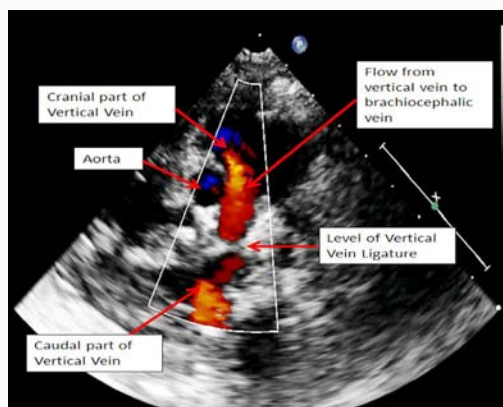


Figure 2: Postoperative transthoracic two-dimensional echocardiogram (day 1 postoperative) with colour flow patterns of venous flow in the unligated vertical vein showing shunting from the left atrium to the brachiocephalic vein via the patent vertical vein.

of the vertical vein ligature resulted in decrease of the pressure to a right ventricle-to-left ventricular pressure ratio of 0.6 and decrease of mean left atrial pressure to 11 mmHg, with a range between 10 and 12 mmHg. This was associated with a significant increase mean arterial blood pressure. The technical details of the adjustable vertical vein ligature have been described in detail in our earlier publication (Figure 1) [6]. The sternum was closed. Pulmonary hypertension was treated with hyperventilation, sedation, phenoxybenzamine, sildanefil and inhaled nitric oxide at 10 to 15 PPM, in varying combination for 120 hours. Postoperatively, the patient was hemodynamically stable on dopamine (7.5µg/kg/min), dobutamine (7.5 µg/kg/min) and milrinone at a dose of 50 µg/kg/IV bolus followed by 0.375-0.75 µg/kg/min. He was sedated and paralysed for first 96 hours. Postoperative echocardiography demonstrated an entirely satisfactory primary repair, with a large anastomosis, larger than the area of the mitral valvular orifice, with no gradient between the pulmonary venous confluence and left atrium and non-turbulent biphasic pulmonary venous flow at <1.2 metres/sec. Serial echocardiography during episodes of pulmonary hypertensive crises demonstrated a left-to-

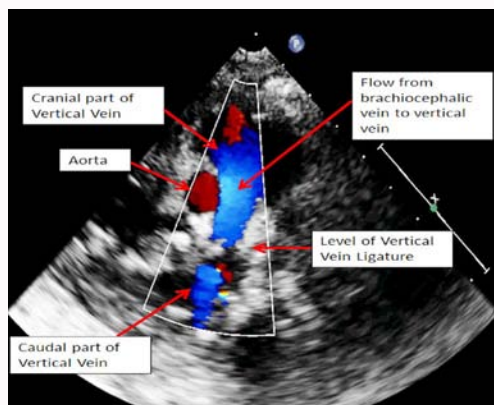


Figure 3: Demonstration of change of the shunting patterns on the 10th postoperative day on the same patient from the brachiocephalic vein to the left atrium via the vertical vein.

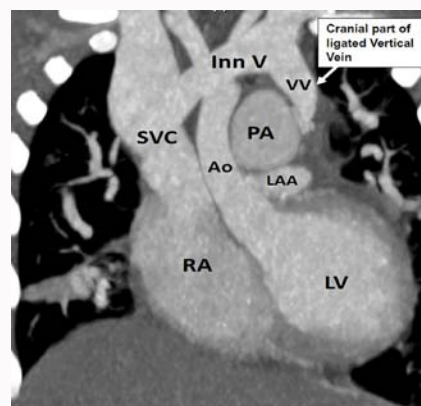


Figure 4: Postoperative postligation, two-dimensional coronal computerized tomographic MIP image showing the cranial part of ligated vertical vein (AO= Ascending Aorta, Inn V= Innominate Vein, LAA= Left Atrial Appendage, LV= Left Ventricle, PA= Pulmonary Artery, RA= Right Atrium, SVC= Superior Vena Cava, VV= Vertical Vein).

right shunting through the patent vertical vein, clearly documenting its role as a temporary vent (Figure 2).

He was extubated on 6th postoperative day with stable hemodynamics. However, he started getting tachypnea and there was evidence of right heart failure between 18-20 days postoperatively. Transthoracic, 2-dimensional, color flow Doppler, echocardiography demonstrated reversal of shunting pattern from the brachiocephalic vein to the left atrium (Figure 3). At this point, we decided to tighten the adjustable ligature in increments. Tightening was achieved by placing additional clips between the polytetrafluoroethylene pledget and the previous clip, while monitoring the left atrial and pulmonary artery pressures 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. The ligature was tightened gradually over a period of 24 to 48 hours, maintaining stable hemodynamics with controlled pulmonary artery pressure at subsystemic levels and normal arterial blood gases. On achieving complete interruption of blood flow through the vertical vein, the ends of the sutures was internalized by making a small skin-incision at the exit points of two sutures. The threads under the skin were secured and marked using a ligaclip individually over each thread.

Another PTFE felt was placed above the marker clip, both sutures were doubly clipped and the sutures were cut flush on the ligaclip to release the portion of the redundant sutures. The skin was closed with two interrupted sutures (Figure 1).

The patient had gradual reduction of cardiomegaly and had an uneventful postoperative course. At 36 months follow-up, the child was asymptomatic showed no clinical evidence of cardiac failure, with Ross's clinical score of 2 and without antifailure cardiac medications.¹⁷ Cross-sectional and Doppler echocardiography revealed normal biventricular function and absence of flow through the atrial septal fenestration and the vertical vein. Follow-up computerized tomographic angiocardiograms demonstrated absence of flow through the vertical vein and there was no distortion of the left superior pulmonary vein and left brachiocephalic veins (Figures 4-6).

Discussion

Reports addressing the issue of not ligating the vertical vein following repair of TAPVC and related postoperative hemodynamics are limited and conflicting.⁵⁻¹⁶ An unligated vertical vein might be a desirable option in patients undergoing rechanneling of supracardiac obstructive Totally Anomalous Pulmonary Venous Connection with post-bypass suprasystemic pulmonary artery pressure in order to decrease perioperative pulmonary artery pressure and pulmonary hypertensive crises [4,6,7,10,11,13]. In 2007, we documented our observations that acute vertical vein ligation resulted in elevated left atrial pressure and impaired left ventricular function suggesting that for a period of time the small, poorly compliant left ventricle of the patient with obstructive Totally Anomalous Pulmonary Venous Connection was unable to maintain adequate cardiac output. Indeed, it is routine to see elevated left atrial pressure and pulmonary artery pressure combined with systemic hypotension and decreased cardiac output after weaning from cardiopulmonary bypass in patients with obstructive totally anomalous pulmonary venous connection.⁴ According to data from the current literature, similar observations have been documented by some investigators [4,7,10,11,13]. On the other hand, in the study reported by Kelle and colleagues, the vertical vein was ligated in all patients with obstructive and non-obstructive Totally Anomalous Pulmonary Venous Connection and postoperative pulmonary hypertension was managed by nitric oxide therapy and conventional ventilator management strategies [18]. Since the pulmonary veins/capillaries do not have any valves, any rise in left atrial pressure will lead to pulmonary hypertension. The postoperative course targets management of these issues and occasionally extracorporeal membrane oxygenation is necessary while the pulmonary vascular bed recovers and left ventricular compliance improves [19,20]. The time course of this adjustment is variable. It is conjectured that there is a feedback loop whereby acute elevation of left atrial pressure results in excessive pulmonary arteriolar constriction with out-of-proportion pulmonary hypertensive response that further exacerbates the low cardiac output state caused by a poorly compliant left ventricle [16]. In order to test our postulates, we embarked on a program of not ligating the vertical vein in selected patients (n=48) undergoing repair of isolated totally anomalous pulmonary venous connection. Only patients with type I, III and IV Totally Anomalous Pulmonary Venous Connection with a discernible ascending or descending vertical vein were included in our previous study [4]. In our previous investigation the following criteria were taken into consideration for maintaining a patent vertical vein:

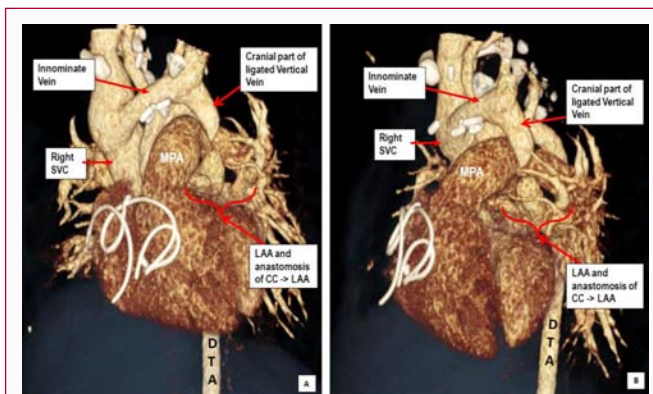


Figure 5: Postoperative post ligation, three-dimensional volume rendered coronal computerized tomographic image- A) anterior, B) anterolateral view showing the cranial part of the ligated vertical vein and the wide unrestricted anastomosis between the common pulmonary venous chamber and left atrial appendage (CC= Common Pulmonary Venous Chamber, DTA= Descending Thoracic Aorta, LAA= Left Atrial Appendage, SVC= Superior Caval Vein).

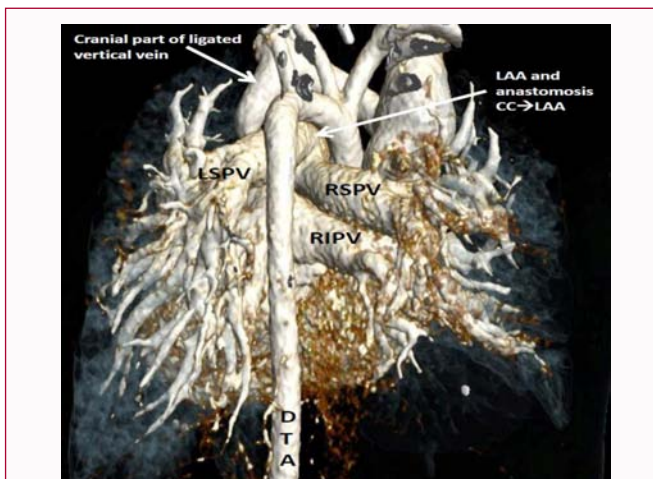


Figure 6: Postoperative post ligation, three-dimensional volume rendered image in posterior projection showing all pulmonary veins draining into the common pulmonary venous chamber which has been anastomosed to the left atrial appendage (CC= Common Pulmonary Venous Chamber, DTA= Descending Thoracic Aorta, LAA= Left Atrial Appendage, LSPV= Left Superior Pulmonary Vein, RIPV= Right Inferior Pulmonary Vein, RSPV= Right Superior Pulmonary Vein).

- Significant elevation of pulmonary arterial or left atrial pressures upon snaring the vertical vein after achieving an adequate unrestricted common pulmonary venous chamber to left atrial anastomosis and left atrial augmentation was considered to be a useful clinical indicator of impaired left atrial compliance and/or disease-related/bypass-related pulmonary vasoreactivity.

- Subsequently, all patients with non-obstructed Totally Anomalous Pulmonary Venous Connection without pulmonary arterial hypertension and patients with obstructed Totally Anomalous Pulmonary Venous Connection with moderate post-bypass pulmonary hypertension, with post-bypass systolic pulmonary arterial pressure from 31 to 50 millimetres of mercury also had a patent vertical vein with an adjustable vertical vein ligature to tide over the postoperative morbidity and mortality.

- All patients with supracardiac Totally Anomalous Pulmonary Venous Connection underwent routine left atrial augmentation using a Dacron polyester fabric and those with

moderate pulmonary arterial hypertension had an elective atrial septal fenestration for right ventricular decompression in the event of perioperative pulmonary hypertensive crisis [6].

In order to allow the left heart to adopt and maintain adequate cardiac output, we have used a redundant Dacron patch for interatrial septum, deviating the same to the enlarged right atrium and have incorporated part of the vertical vein to achieve structural alignment and augmentation of the left atrial cavity [4,6]. The concept of enlarging the left atrium 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 a Dacron patch graft to close the atrial septal defect and enlarge the left atrium [21,22].

In this patient, the decision to keep the vertical vein patent was made after the occurrence of post-bypass systemic or supra-systemic pulmonary artery pressure upon snaring the vertical vein. In the absence of anastomotic stricture, this undesirable effect can be explained by decreased unloading of the pulmonary venous chamber due to non-compliant left-sided chambers. Incorporation of the vertical vein augmented the left atrium, served to lower pulmonary arterial and vertical vein pressure almost immediately and acted as a “pop-off” valve during episodes of pulmonary hypertensive crises, lessened duration of ventilation and inotropic support and improved short-term and long-term survival by providing superior hemodynamics (Figure 2).

In an attempt to permit the “spill over” or pop off in the setting postoperative right ventricular dysfunction, our group adopted the practice of atrial septal fenestration in patients with obstructive supracardiac and infracardiac totally anomalous pulmonary venous connection. During episodes of pulmonary hypertensive crises with limited right ventricular output and an elevated central venous pressure, 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 [4,6]. Thus, the unligated vertical vein in conjunction with a calibrated atrial septal fenestration, decompressed the small left atrium after repair, equalized the left atrium and central venous pressure and was the automatic choice to avoid a dismal outcome in the perioperative period. There is no consensus in the published literature regarding the fate of unligated vertical vein following repair of totally anomalous pulmonary venous connection [5-15]. Literature documents clear anecdotal cases of spontaneous involution of the anomalous vertical vein at one end of the spectrum and a functioning conduit with shunt induced cardiac failure at the other end [5-15]. Spontaneous closure of an unligated vertical vein is contingent upon normal growth and function of left cardiac chambers [7].

Between 1997 and 2006, 27 out of 48 (46.5%) patients did not undergo vertical vein ligation [4]. Contrary to the reports by Cope and colleagues in which patent vertical vein atrophied, 11 of 23 survivors in our study had symptoms of a large left-to-right shunt through the unligated vertical vein requiring delayed closure in all cases. The vertical vein was ligated through re sternotomy in 4 patients, left anterolateral thoracotomy in 2 cases and adjustable vertical vein ligation in 5 patients. Although delayed closure of the vertical vein was successful in all cases, with concomitant elevation of pulmonary artery pressure, it was attended by extremely high left atrial pressure in six patients and proved a difficult postoperative challenge. These findings were suggestive of a relatively small, non-compliant dysfunctional left-sided chamber or of disease-related or

cardiopulmonary bypass-related pulmonary vasoreactivity [4].

Based on the literature and reasoning cited above, the authors embarked on a programme of routine adjustable vertical vein ligation in patients with obstructed Totally Anomalous Pulmonary Venous Connection with post-bypass systemic or suprasystemic pulmonary arterial hypertension and published the initial observations on this novel device for ligation of the vertical vein in Cardiology in the Young [6]. Whether an adjustable vertical vein ligature is advantageous over routine vertical vein ligation has remained debatable. Clinical studies on an unligated vertical vein in the setting of obstructive supracardiac Totally Anomalous Pulmonary Venous Connection are too limited and insufficient to generate evidence-based guidelines.

A persistent left-to-right shunt with right heart 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 transcatheter vertical vein closure [6,23]. An adjustable vertical vein ligature allows easy tightening in increments with gradual increase in ventricular after load without causing unstable hemodynamics under optimal physiological conditions once the disease-related / bypass-related pulmonary vaso-reactivity disappears without re sternotomy or thoracotomy.

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 PTFE 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 (Figure 1). Postoperatively, we performed computed tomographic angiography on this patient at follow-up and discovered no untoward findings (Figures 4-6).

Conclusions

We conclude that an unligated vertical vein in a subset of patients with obstructed TAPVC and supra-systemic post-bypass pulmonary hypertension facilitates reduction of pulmonary artery pressures, thus avoiding pulmonary hypertensive crises and postoperative low cardiac output syndrome and contributes to a favourable outcome.

The routine use of a percutaneously adjustable vertical vein ligature in patients with obstructed supracardiac TAPVC with a discernible vertical vein and pulmonary hypertension allows gradual tightening or loosening of the ligature under optimal physiologic conditions without reoperation and without causing any distortion of the left superior pulmonary vein and left brachiocephalic vein.

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