



The Vertical Vein in Patients with Obstructive Supracardiac Totally Anomalous Pulmonary Venous Connection: we can ligate it but should we ligate it?

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Abstract

The present perspective is a synthesis of all published literature on selective vertical vein patency and adjustable vertical vein ligation in the setting of rechanneling of obstructed supracardiac totally anomalous pulmonary venous connection (TAPVC) in order to decrease the episodes of perioperative pulmonary hypertensive crises, postoperative low cardiac output syndrome and mortality. Additionally, this manuscript attempts to address the guidelines for candidate selection for selective vertical vein patency in patients with obstructive supracardiac TAPVC.

Keywords: Vertical vein; Totally anomalous pulmonary venous connection; Pulmonary hypertensive crises; Adjustable vertical vein ligation

Perspectives on the Vertical Vein Ligation in TAPVC

The medical literature is rife with examples of therapies that seemed likely to be beneficial, but were in fact either pointless or harmful. Traditionally, ligation of the vertical vein at the time of rechanneling of totally anomalous pulmonary venous connection (TAPVC) has been recommended to prevent the perceived consequences of a residual left-to-right shunt [1-6]. As yet, there are no specific criteria for candidate selection to maintain a patent vertical vein and there is no consensus in the literature regarding the fate of unligated vertical vein following repair of TAPVC. 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 [1-10].

Despite improvements in pediatric anaesthesia, intensive care, diagnostic accuracy with echocardiography and computerized angiography, often obviating the need for cardiac catheterization and advances in surgical techniques during the past decade, repair of obstructive TAPVC continues to be associated with mortality ranging from 10 to 50 percent [1-13].

Careful analysis of the published literature substantiates significant incidence of low cardiac output following rechanneling of obstructed TAPVC [1-13]. We certainly know that recurrent episodes of pulmonary hypertensive crises secondary to disease-related/cardiopulmonary bypass (CPB)-related heightened pulmonary vasoreactivity, rapid development of pulmonary vascular medial hypertrophy, a small non-compliant left atrium (LA) and left ventricle (LV) have been variously implicated as the causative factors for low cardiac output syndrome (LCOS). We also know about the worse outcomes of TAPVC repair due to delayed referral, accounting for cardiac cachexia, emergency operation, intrinsic pulmonary venous obstruction, pulmonary infection and severe pulmonary hypertension [1-13].

Reports of left heart volume characteristics in obstructive TAPVC have varied considerably, from normal findings to the conclusion that the left sided chambers are smaller than normal and the LA lacks both normal compliance and reservoir function [13-23]. Such chamber abnormalities have been attributed to large left-to-right shunt causing reduced atrial filling and decreased left ventricular relaxation secondary to elevated right ventricular diastolic pressure or volume [13-23]. However, we do not know what percentage of patients with obstructive TAPVC have structurally smaller left-sided chambers and a non-compliant and dysfunctional LV compared to non-obstructive cases [1-23].

We have known for a while that creation of a large, tension-free anastomosis, precise geometric

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alignment of the pulmonary venous chamber with the body of LA avoiding torsion and rotation of pulmonary veins, introduction of phenoxybenzamine in the management of pulmonary hypertensive crisis and delayed sternal closure are the factors for reduction of perioperative mortality after repair of TAPVC in recent years [1,2,9-13]. Future publications auditing the above-mentioned issues will add nothing to our understanding of the problem of postoperative LCOS. We need to identify the anatomic and physiologic issues involved in a subset of patients with obstructive TAPVC. The issues to be resolved are: a) identification of the structurally smaller left sided chambers, b) identification of non-compliant and dysfunctional LV, c) identification of concomitant disproportionately increased pulmonary vascular medial thickness, and finally, d) identification of concomitant hypoplasia of the pulmonary venous system.

In 2007, the authors documented their observations that acute vertical vein ligation resulted in elevated LA pressure and left ventricular function and cardiac output are negatively impacted 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 [21]. Indeed, it is routine to see elevated LA pressure and pulmonary artery pressure combined with systemic hypotension and decreased cardiac output after weaning from CPB in patients with obstructive TAPVC [21,22]. 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. The time course of this adjustment is variable. It is conjectured that there is a feedback loop whereby acute elevation of LA 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 [22].

In order to test the above-mentioned postulates to reduce the perioperative mortality and morbidity, the authors embarked on a program of routine adjustable vertical vein ligation in patients with obstructed TAPVC with post bypass systemic or suprasystemic pulmonary arterial hypertension [21-23]. Thus, there were seven forces driving our criteria for selection of patients whose vertical vein was kept patent after rechanneling of TAPVC:

- i. The desire to reduce the pulmonary artery pressure in the perioperative period after achieving an adequate-sized, unrestrictive anastomosis along with pharmacological manipulations.
- ii. The desire to reduce pulmonary hypertensive crises, low cardiac output, and hospital mortality following repair of TAPVC with pulmonary hypertension.
- iii. The desire that the unligated vertical vein 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.
- iv. The desire that the unligated vertical vein may 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.
- v. The desire to gradually tighten or loosen the ligature under optimal physiologic conditions, once the disease-related/bypass-

related pulmonary vaso-reactivity disappears.

vi. The desire to gradually increase the ventricular afterload without causing unstable hemodynamics under optimal physiologic conditions, and

vii. The desire to occlude the vertical vein in the event of a significant left-to-right shunt and right heart failure without re-sternotomy or thoracotomy.

An initial concern about this technique was the possibility of distortion of the left upper pulmonary vein and left brachiocephalic vein. To address these concerns, we have performed the following manoeuvres: (i) we threaded the loop ligature through a polytetrafluoroethylene felt and secured the same with the adventitia of the vertical vein to prevent its displacement, and (ii) both the 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 straight lie, avoiding subsequent distortion or occlusion of the left superior pulmonary and brachiocephalic veins [23].

In our initial investigations on 48 patients undergoing rechanneling of TAPVC between 1997 and 2006, 27 (46.5%) patients did not undergo vertical vein ligation [21]. Contrary to the report by Cope and colleagues, in which patent venous pathway atrophied, 11 of 23 survivors of obstructive supracardiac TAPVC allowed symptoms of a large left to right shunt through the unligated vertical vein requiring delayed closure of the vertical vein in all cases [21]. 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 relatively small, non-compliant, dysfunctional left-sided chambers or of disease-related or CPB-related pulmonary vasoreactivity [21].

Subsequently, in 2007 we introduced the concept of adjustable vertical vein ligation in the setting of obstructive supracardiac TAPVC with the aims and objectives as narrated above [23]. We demonstrated that the unligated vertical vein during repair of obstructed TAPVC is associated with decreased episodes of pulmonary hypertensive crisis, postoperative low cardiac output syndrome, and lessened duration of ventilation and inotropic support, provided early normalization of hemodynamics and decreased in hospital mortality. There were no late deaths. At a mean follow-up of 33.34 ± 29.88 months, median 30 months, the actuarial survivals were $92.6\% \pm 0.05\%$ in the unligated category and $71\% \pm 0.08\%$ for the ligated category ($p=0.03$). Since our last publications in 2007, an additional 115 patients with supracardiac TAPVC underwent adjustable vertical vein ligation, rechanneling of supracardiac TAPVC, routine left atrial augmentation and a patent shunt at the atrial level. The conclusions based on the original 48 patients remain valid. All patients underwent serial cross-sectional and Doppler echocardiographic evaluation in the postoperative period and gradual process of vertical vein ligation at varying time intervals between 5-25 days, as soon as right to left shunting through the vertical vein disappeared. None required anti-failure cardiac medications. Follow-up computed tomographic angiocardiograms in all patients demonstrated absence of flow through the vertical vein and ruled out distortion at the left upper pulmonary vein and left brachiocephalic veins [21-23].

Whether an adjustable vertical vein ligation with concomitant

rechanneling of supracardiac TAPVC is advantageous over the traditional concept of routine vertical vein ligation is a subject of debate. Since all investigators and surgeons have not accepted these findings or utilized these techniques, the answer to the above postulates and observations is forthcoming.

It is pertinent to state that a persistent left-to-right shunt and 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 ligation or percutaneous angiographic vertical vein embolization.

Additionally, in all cases of obstructed and non-obstructed supracardiac TAPVC, we introduced the following modifications:

- i. All patients underwent rechanneling of supracardiac TAPVC via the posterior approach, dislocating the apex of the heart into right pleural cavity, achieving a wide, non-restrictive anastomosis without any geometric alteration.
- ii. All patients underwent routine LA augmentation via right atriotomy using a dacron patch. This served not only to enhance the capacity of the LA but also ensured adequate-sized communication between the pulmonary veins and left atrium.
- iii. All patients underwent percutaneously adjustable vertical vein ligation and gradual occlusion one to two weeks after extubation ensuring no right-to-left shunting through the vertical vein.
- iv. All patients with obstructed TAPVC underwent fenestration of the atrial septal patch in addition [21-23].

The mechanisms causing heightened pulmonary vasoreactivity following repair of TAPVC are multifactorial, and may reflect release of platelet-activating factors, endothelin, and arachidonic acid metabolites from pulmonary endothelial cells, decreased ratio of prostacyclin to thromboxane and a decline or absence of acetylcholine responsiveness [1-13,24,25]. Many agents have been advanced as being optimal on the grounds that they selectively reduce pulmonary vascular resistance, but few do so, and no clearly superior one has been identified. Therefore, in the authors' centre, a varying combination of fentanyl, hyperventilation, correction of acidosis, inhaled nitric oxide, sodium nitroprusside, and phenoxybenzamine were used to manage pulmonary hypertensive crises [21,23].

Traditionally, an intentional atrial septal fenestration is created permitting the "spill-over" or "pop-off" in the setting of postoperative right ventricular dysfunction following intracardiac repair of tetralogy of Fallot, pulmonary atresia, Ebstein's anomaly, Rastelli's operation and one and one-half ventricular repair [21-23]. A review of the literature on this topic reveals that it can be performed relatively safely and patients do reasonably well at follow-up, albeit at the expense of mild desaturation.

James S. Tweddell in his editorial on the issue of vertical vein ligation revisited some observations during the early years of cardiology and cardiac intervention [22]. In 1916, Lutembacher described the impact of an atrial septal defect on acquired mitral stenosis by trading mild elevation of right atrial pressure and pulmonary over circulation for acute elevation of LA pressure [22]. In 1949, Bland and Sweet anastomosed azygous vein to pulmonary vein to improve the functional status of patients with critical rheumatic mitral stenosis permitting spill over of the congested pulmonary veins

into the capacious systemic venous system [22]. Following Bland and Sweet's experiment, future efforts were directed at the mitral valve itself and heralded the beginning of cardiac surgery [22]. Revisiting these observations of Lutembacher, Bland, Cope, and Chowdhury suggest that the systemic venous system and pulmonary vasculature can accommodate the excess volume and limit the impact of excessive LA pressure. Eventually, with continued shunting in the event of uncorrected primary problem, the pulmonary vasculature and the systemic venous circuit will be overwhelmed and right sided cardiac failure will develop [21-23].

Since 2000, it has been the author's practice to perform atrial septal fenestration in patients with obstructive supracardiac and infracardiac TAPVC. 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 [21-23].

The decision to keep the vertical vein patent is made after the occurrence of post-bypass systemic or supra-systemic PA 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 LA, served to lower pulmonary arterial and vertical vein pressure almost immediately and acted as a "pop-off" valve to the systemic venous system during episodes of pulmonary hypertensive crises [21-23]. Thus, the unligated vertical vein in conjunction with a calibrated atrial septal fenestration, decompressed the small LA after repair, equalized the LA and central venous pressure and was the automatic choice to avoid a dismal outcome in the perioperative period [21,23].

In our previous study, we demonstrated that a patent vertical vein during repair of obstructed TAPVC reduced pulmonary artery pressure, decreased perioperative hypertensive crisis, provided a temporary vent during pulmonary hypertensive crisis and improved short-term and long-term survival by providing superior hemodynamics [21,22]. Even then, the postoperative course of a subset of patients with obstructed TAPVC and very small individual pulmonary veins is complex and the prognosis is poor [1,2,11,14,24,25]. Spontaneous closure of an unligated vertical vein is contingent upon normal growth and function of left cardiac chamber [7,9,10].

Subsequently, we embarked on a program of routine deployment of an adjustable vertical vein ligation in all patients with obstructive supracardiac TAPVC with more than moderate post CPB pulmonary hypertension [21-23]. This strategy allowed a more gradual occlusion of the vertical vein so that acute elevation of the LA pressure was avoided, albeit at the expense of some period of pulmonary over circulation and elevated right-sided filling pressure, thereby tiding over the post operative phase of heightened pulmonary vasoreactivity. Such a band allowed easy tightening in increments, with gradual increase of ventricular after load without the need for multiple reoperations [21-23].

Autopsy findings (n=4) revealed a small pulmonary venous confluence, diffuse hypoplasia, intimal hypertrophy, increased pulmonary vascular medial thickness, pulmonary lymphangiectasia and interstitial emphysema [21]. Anatomic studies by other investigators have shown that preoperative pulmonary venous

obstruction is associated with increased medial thickness of pulmonary vasculature that disproportionately exceeds the degree of change observed in other lesions with left-to-right shunt induced pulmonary hypertension [21,24]. On the basis of these observations, we speculate that the medial and intimal changes seen in preoperative obstruction may predispose towards the development of intrinsic pulmonary vein stenosis. We concur with the observations of other investigators that an unligated vertical vein in this subset of patients with co-existing pulmonary arteriopathy exerts an unfavorable effect on the morbidity and surgical outcome despite adequate pulmonary venous decompression [21,24]. Given the bleak prognosis for these patients, alternative management strategies like lung transplantation may perhaps be considered [25].

With these objectives, in order to reduce the perioperative mortality and morbidity, we attempted to evaluate outcomes following selective use of vertical vein ligation in a consecutive series of patients undergoing repair of isolated TAPVC. Only patients with types I, III or IV TAPVC with a discernible ascending or descending vertical vein were included in this study [21-23].

What we do not know about Obstructive TAPVC and How can we acquire the Information Necessary to Formulate these Criteria?

Acquiring the information that documents a patent vertical vein is beneficial relative to the alternative operation of ligation in the setting of obstructive TAPVC is more difficult, yet it is badly needed. One place to begin would be to define a large population of patients with obstructive TAPVC undergoing rechanneling and assign them to one of the two categories in the presence of moderate-to-severe pulmonary arterial hypertension after coming off bypass. The unligated category could then be compared to the ligated category. A careful quantitative evaluation of the preoperative morphologic and physiologic characteristics of both left and right-sided chambers for all patients with obstructive TAPVC could then be used to determine whether and under what left heart conditions, the unligated vertical vein is the more beneficial option. Such information would be welcome and noteworthy. Nothing in the literature even remotely addresses the issue of making a quantitative assessment of the left side of the heart that can be used to objectively decide between the surgical options. The surgeon is left to make a clinical (subjective) decision between the two operations.

To properly test the hypothesis that “vertical vein ligation results in inferior outcome and non-ligation in selected subsets of TAPVC results in superior outcome”, a multi-institutional, prospective randomized trial of ligation vs. non-ligation would be necessary and would be the last refuge for those who cannot accept the conflicting complex findings of the anatomy and pathophysiology of obstructive TAPVC. Furthermore, the rarity and spectrum of TAPVC would make such a trial difficult.

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