Rechanneling of total anomalous pulmonary venous connection with or without vertical vein ligation: Results and guidelines for candidate selection

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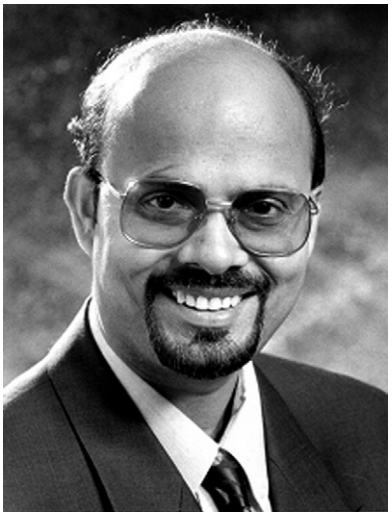

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Supplemental material is available online.

Objective: This study investigated whether postoperative low cardiac output and mortality in obstructed total anomalous pulmonary venous connection could be reduced by selective vertical vein patency.

Methods: Fifty-eight patients undergoing rechanneling of total anomalous pulmonary venous connection between 1997 and 2006 were studied. The vertical vein was left patent in 27 patients (group I) and ligated in 31 (group II). Mean ages were 1.49 ± 1.63 and 4.37 ± 3.38 months for groups I and II, respectively.

Results: Operative mortalities were 29.1% and 7.4% for ligated and unligated groups, respectively (relative risk 1.75, 1.16-2.64, $P = .036$). Age younger than 1 month, obstructive total anomalous pulmonary venous connection, hypoplastic pulmonary veins, pulmonary hypertensive crisis, low cardiac output, and vertical vein ligation were significant risk factors for death according to logistic regression analysis. Patients with obstructed total anomalous pulmonary venous connection undergoing vertical vein ligation demonstrated predominant right ventricular dysfunction (relative risk 2.93, 1.28-6.73, $P = .011$), pulmonary hypertensive crisis (relative risk 2.90, 1.25-6.75, $P = .013$), and 3.28 times the risk of death (95% confidence interval 1.08-9.99, $P = .032$) relative to the unligated group.

Conclusions: In a subset of patients with obstructed total anomalous pulmonary venous connection, an unligated vertical vein reduces pulmonary arterial pressure, decreases perioperative pulmonary hypertensive crises, provides a temporary pop-off valve during pulmonary hypertensive crisis, and improves survival by providing superior hemodynamics. The high mortality in the ligated group suggests that patients with obstructed total anomalous pulmonary venous connection with post-bypass moderate pulmonary hypertension possibly should not undergo vertical vein ligation. We propose routine use of an adjustable ligature around the vertical vein in all patients with more than moderate post-bypass pulmonary hypertension, allowing gradual tightening in increments without multiple reoperations.

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Abbreviations and Acronyms

ASD = atrial septal defect
CI = confidence interval
CPB = cardiopulmonary bypass
LA = left atrium
PA = pulmonary artery
RR = relative risk
SPAP = systolic pulmonary arterial pressure
TAPVC = total anomalous pulmonary venous connection

Despite improvements during the past decade in pediatric anesthesia, intensive care, and myocardial protection, repair of obstructive total anomalous pulmonary venous connection (TAPVC) continues to be associated with significant mortality, reportedly ranging from 10% to 50%.[1-5] In 1999, we reported worse outcomes of TAPVC repair as a result of delayed referral, accounting for cardiac cachexia, emergency operation, pulmonary infection, and severe pulmonary artery (PA) hypertension.[5]

Studies have documented that in the presence of obstructive pulmonary venous drainage, the left-sided chambers are smaller than those in nonobstructive cases.[6-9] Despite innovative surgical techniques to increase the dimension of the left atrial (LA) cavity and pharmacologic manipulations to combat pulmonary hypertensive crisis, low cardiac output remains a significant problem, leading to hemodynamic instability and morbidity after surgery.[1-9]

From these observations, it has been hypothesized that a patent vertical vein may function as a temporary reservoir for pulmonary venous blood after TAPVC repair, volume unloading the small, noncompliant, left-sided cardiac chambers until they are able to grow and adapt to the increased flow demands.[8-11] Not all investigators and surgeons have accepted these findings or used these techniques. To investigate this hypothesis, this study aimed first to ascertain whether the practice of routine ligation of vertical vein is necessary, second to examine the effects of selective patent vertical vein on postoperative hemodynamics, third to compare the outcomes of patients undergoing interruption of the vertical vein with those in whom it was left unligated, and fourth to determine the long-term fate of the unligated vertical vein.

Patients and Methods

Criteria for Decision Making and Selection of Patients

This prospective study evaluated outcomes after selective use of vertical vein ligation in a consecutive series of patients undergoing repair of isolated TAPVC. Only patients with type I, III, or IV TAPVC with a discernible ascending or descending vertical vein were included in this study. The decision to keep the vertical vein patent was made after the occurrence of elevated PA pressure (systemic or suprasystemic) after coming off cardiopulmonary bypass (CPB) and untoward hemodynamic effects on snaring the vertical vein. All patients with nonobstructed TAPVC without pulmonary hypertension and patients with obstructed TAPVC with moderate post-CPB pulmonary hypertension (systolic PA pressure [SPAP] 31-50 mm Hg) underwent vertical vein ligation. Thus, there were four forces driving our criteria for selection of patients in whom the vertical vein was kept patent after rechanneling of TAPVC: (1) the desire to reduce the PA pressure in the perioperative period after achievement of an adequately sized, unrestrictive anastomosis along with pharmacologic manipulations; (2) the desire to reduce pulmonary hypertensive crises, low cardiac output, and in-hospital mortality after repair of TAPVC with pulmonary hypertension; (3) the desire that the unligated vertical vein serve as a temporary pop-off valve in the event of pulmonary hypertensive crises; and (4) the desire that the unligated vertical vein function as a temporary venous reservoir for pulmonary venous blood, volume unloading the small, noncompliant left-sided cardiac chambers until they were able to grow and adapt to the requisite flow demands.

We excluded all patients with anomalous pulmonary venous drainage to the right superior vena cava, coronary sinus, or right atrium who lacked a discernible vertical vein (n = 5). We also excluded children with associated complex lesions, such as atrioventricular canal, transposition of the great arteries, or functionally univentricular heart (n = 3).

Demographics and Preoperative Evaluation

To test our postulates, we embarked on a program of not ligating the vertical vein in selected patients undergoing TAPVC repair at our institution. All patients in this study population were operated on by a single surgeon (U.K.C.), making uniformity in the surgical protocol possible. The patients were entered in the study protocol after informed consent had been obtained from their parents or guardians. Fifty-eight consecutive patients undergoing rechanneling of isolated TAPVC from January 1997 through March 2006 at All India Institute of Medical Sciences, New Delhi, were included in this prospective study (Figure E1). In this study, 27 patients (46.5%) underwent rechanneling of TAPVC without vertical vein ligation (group I), and 31 patients (53.5%) underwent vertical vein ligation (group II). Their demographic and clinical profiles are depicted in Table 1.

Patient age at operation ranged from 1 day to 8 months (mean 1.49 ± 1.63 months, median 1 month) among those who did not undergo vertical vein ligation and from 1 to 12 months (mean 4.37 ± 3.37 months, median 3 months) among those who did. Fourteen patients (24.1%) were younger than 1 month, and 9 (15.5%) patients were between 6 and 12 months (Figure E2). Because of the uses of selective ligation, there were significant differences in the baseline characteristics of the two groups (Table 1). Most patients in this study group were small for age and 53.5% weighed less than the 50th percentile of predicted weight for Indian neonates and infants.

Echocardiography was our principal diagnostic modality. Transthoracic 2-dimensional, color flow, Doppler echocardiography was performed (Sonos 5500; Hewlett-Packard Company, Palo Alto, Calif), and patients were categorized according to the site of pulmonary venous drainage. Cardiac catheterization and angiocar-

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