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Perioperative and Anesthetic Considerations in Total Anomalous Pulmonary Venous Connection

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is a potentially devastating form of congenital heart disease in which all pulmonary blood flow returns to the systemic venous circulation rather than the left atrium. Anomalous pulmonary venous flow may be obstructed at birth, and affected infants present with severe cyanosis and poor cardiac output unresponsive to standard resuscitation with prostaglandin. Obstructed TAPVC remains one of the few indications for emergent neonatal cardiac surgery. This review will discuss the physiology and perioperative management of isolated TAPVC without associated cardiac lesions.

Keywords

total anomalous pulmonary venous return, congenital heart disease, cardiac anesthesia, intraoperative assessment, postoperative complications

Introduction

Total anomalous pulmonary venous connection (TAPVC) is a relatively uncommon type of congenital heart disease, representing less than 3% of congenital cardiac anomalies.[1-5] Approximately 35% of children have complex TAPVC with coexisting cardiac lesions, including heterotaxy, single ventricle, and complete atrioventricular canal defect.[1,3-7] This review will focus only on isolated TAPVC without other cardiac lesions, apart from patent ductus arteriosus (PDA), atrial septal defect (ASD), or patent foramen ovale (PFO).

Embryology, Anatomy, and Classification

TAPVC results from failure of normal fetal involution of the connection between the pulmonary venous plexus and the systemic veins, leaving the primitive connections intact.[1,6,7] During normal development, the common pulmonary vein begins as an outgrowth of the dorsal atrial wall and connects to the pulmonary venous plexus, which communicates with the cardinal and umbilicovitelline systems. The primitive connections between the pulmonary and systemic circulation typically involute as the common pulmonary vein is incorporated into the left atrium. Disruption of this process results in failure of the connection between the pulmonary venous confluence and the atrium, leaving the venous confluence in connection with the systemic venous system at the level of the heart or above or below the diaphragm.[1,3-9]

The location of the connection of the pulmonary veins to the systemic circulation varies: The connection is supracardiac in about 45% of cases, cardiac in 25%, infracardiac in 20%, and mixed in 10%.[4,6,8-11] In supracardiac TAPVC the pulmonary veins typically drain to a confluence behind the left atrium, which is connected via an ascending vertical vein to the left innominate vein. Less commonly the vertical vein drains to the superior vena cava (SVC) or the azygous vein (Figure 1). In cardiac TAPVC, the veins drain to the coronary sinus or directly into the right atrium (Figure 2). In infracardiac TAPVC, the vertical vein most frequently descends to join the portal vein or the ductus venosus.[4,6,7,11-13] Connection to the gastric or hepatic veins or directly to the inferior vena cava have also been reported (Figure 3).[6,7,12-14] Nearly half of mixed TAPVC cases are characterized by an asymmetrical ("3 + 1") connection in which 3 of the pulmonary veins drain to a common site via

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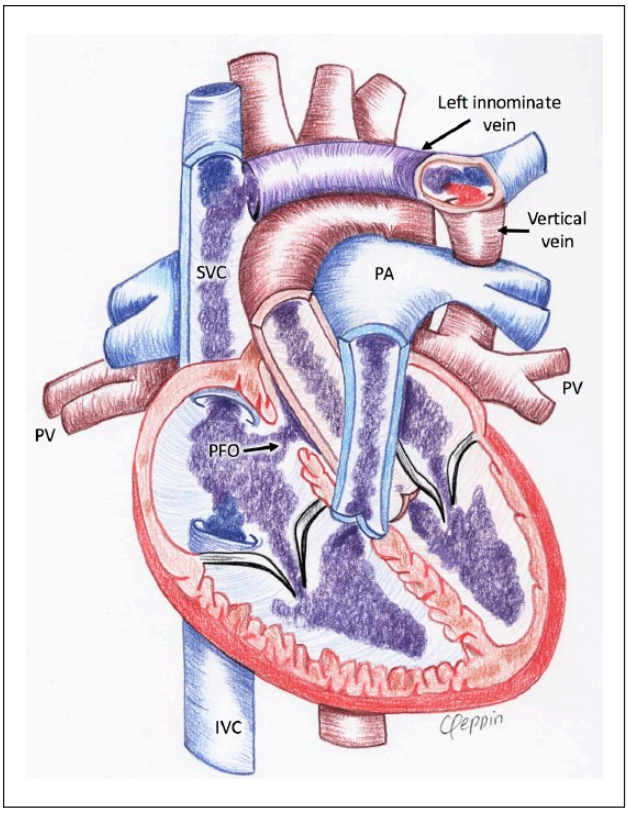

Figure 1. Supracardiac total anomalous pulmonary venous connection (TAPVC) with connection of the pulmonary venous confluence via a vertical vein to the left innominate vein.

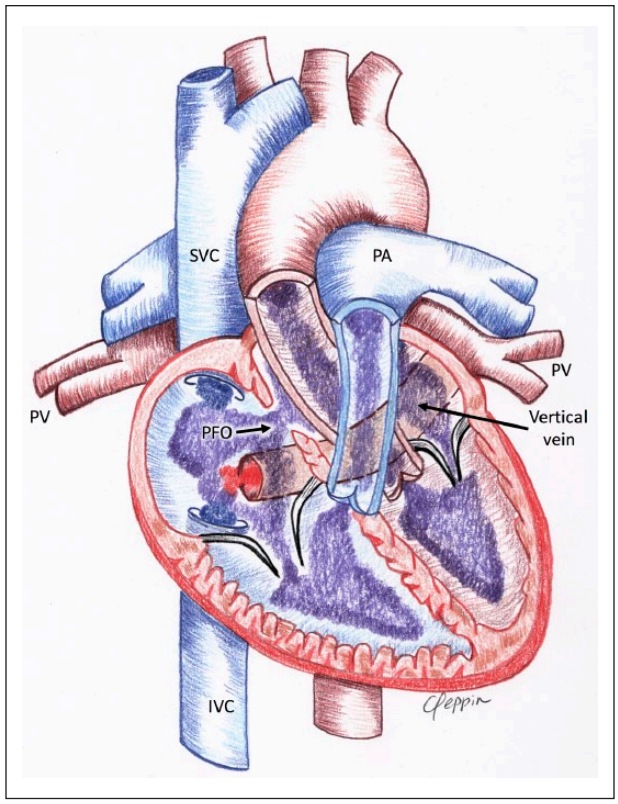

Figure 2. Cardiac total anomalous pulmonary venous connection (TAPVC) with connection of the pulmonary venous confluence to the coronary sinus.

a confluence and the remaining pulmonary vein drains to a separate site. Bilateral, symmetric connections with separate drainage of the veins from the right and left lungs ("2 + 2") are also frequently described. Other anatomic variants, such as individual drainage of all 4 veins, are less common.[14,15] The pulmonary veins in mixed TAPVC have been reported to connect to the right atrium or nearly any vein in the upper thorax and mediastinum.[15]

Greater than 30% of cases of TAPVC present with pulmonary venous obstruction.[1,4,5,11,15] Obstruction is nearly universal in neonates with infracardiac TAPVC and uncommon in cardiac TAPVC.[1,4-6,11,15] Supracardiac TAPVC is obstructed in approximately 50% of children.[1,5,6,11] The site of obstruction in supracardiac TAPVC may be at the junction of the pulmonary venous confluence and the vertical vein, the junction of the vertical vein with the innominate vein, or the innominate vein to the SVC. If the vertical vein runs posterior to the left pulmonary artery (LPA), it can become compressed between the LPA and the bronchus, creating a vicious cycle where increasing PA pressure leads to worsening obstruction. In infracardiac TAPVC, obstruction generally occurs at the diaphragm or the ductus venosus.[3,5,6,11] Obstruction due to closure of the ductus venosus causes acute and severe instability, whereas obstruction due to sluggish pulmonary venous blood flow through the portal venous system and the hepatic capillary bed presents more insidiously.[6]

Natural History and Physiology

TAPVC is often diagnosed postnatally given the challenge of identifying the subtle echo findings related to limited pulmonary blood flow in utero.[16,17] The timing and severity of presentation is primarily dependent on the degree of obstruction of pulmonary venous return and restriction of right to left shunt across the atrial septum.

The majority of children with unobstructed TAPVC are symptomatic in infancy and eventually present with right ventricular volume overload and right heart failure. A small minority of patients are relatively asymptomatic into adulthood.[4,18-20] In the absence of pulmonary venous obstruction, the anomalous venous return imposes a volume load on the right heart resulting in right atrial and right ventricular dilation.[6] Right to left shunting across the atrial septum results in cyanosis, and any restriction at the atrial shunt limits left ventricular (LV) preload. Increased pulmonary blood flow eventually leads to muscularization of the pulmonary vascular bed and may lead to the

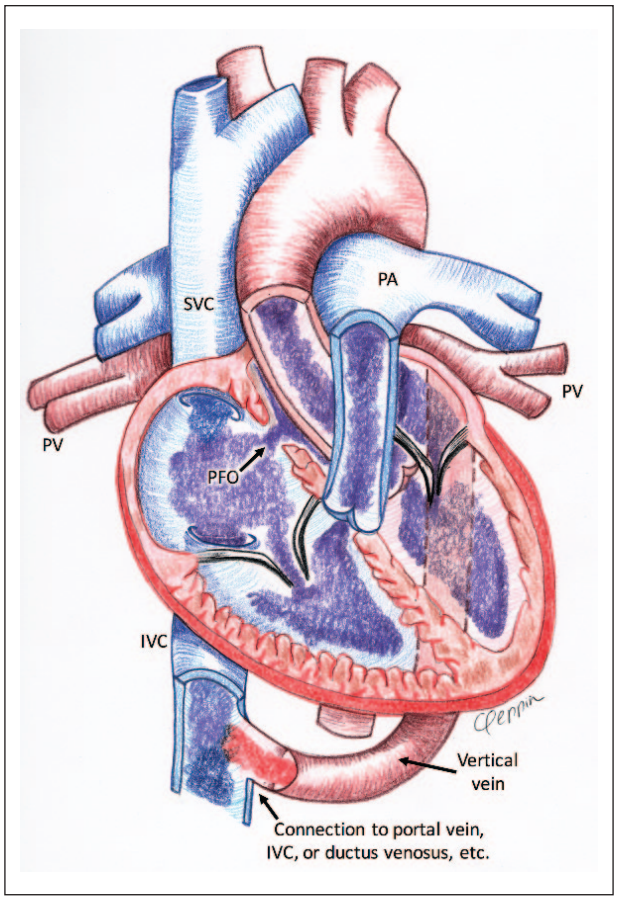

Figure 3. Infracardiac total anomalous pulmonary venous connection (TAPVC) with connection of the pulmonary venous confluence via a vertical vein to the inferior vena cava.

development of irreversible pulmonary hypertension.[13] Although overall left ventricular mass is normal in patients with TAPVC, right ventricular pressure and volume overload accompanied by diminished LV filling cause leftward displacement of the interventricular septum and distortion of LV architecture. The interventricular septum is often hypertrophied with abnormal orientation of myocardial fibers, despite a normal to reduced LV free wall thickness. Such changes to the LV can become irreversible if correction is delayed.[13,18,21,22]

Neonates with obstructed pulmonary venous flow present with profound cyanosis and acidosis shortly after birth.[23] These patients have a bidirectional cardiac shunt. A left to right shunt occurs as the oxygenated blood from the pulmonary veins enters the systemic venous system. The obligatory atrial communication provides a right to left shunt of mixed pulmonary and systemic venous blood. This results in cyanosis that is unresponsive to increased FiO_2 and prostaglandin (PGE).[6] Occasionally, neonates with obstructed TAPVC deteriorate with the introduction of PGE; increasing ductal flow in the face of fixed pulmonary venous obstruction can result in severe pulmonary edema and clinical decompensation.[24]

Because mixing of oxygenated and deoxygenated blood occurs at the systemic venous or right atrial level, saturation should be the same in all chambers of the heart. Pre- and postductal saturations are typically similar; however, in the case of infracardiac TAPVC with a patent ductus, preferential streaming of deoxygenated SVC blood through the tricuspid valve to the right ventricle (RV), PA, ductus, and descending aorta and streaming of oxygenated inferior vena cava blood to the left atrium, LV, and ascending aorta can result in a higher oxygen saturation in the upper extremities than the lower extremity saturation (differential cyanosis). In supracardiac TAPVC, the opposite situation can occur, leading to higher saturation in the lower extremities than the upper extremities (reverse differential cyanosis).[25]

Pulmonary vascular changes are observed shortly after birth in cases of obstructed TAPVC.[13] Autopsies of deceased infants with obstructed TAPVC demonstrate increased arterial medial thickness, intimal proliferation in preacinar veins, and abnormally small and thick-walled extrapulmonary veins. These changes leave neonates extraordinarily vulnerable to pulmonary hypertension in the perioperative period and may predispose patients to development of pulmonary venous obstruction despite successful repair.[26]

Preoperative Evaluation

Preoperative evaluation should begin with a thorough history and physical examination. Newborns with obstructed veins are generally intubated and critically ill, whereas patients without obstruction may present with milder cyanosis and dyspnea resulting from pulmonary overcirculation. As noted above, pre- and postductal saturations are generally similar.

Electrocardiogram (ECG) displays evidence of RV hypertrophy, and chest radiograph often reveals bilateral pulmonary venous congestion.[8,13,15] Asymmetric pulmonary venous congestion suggests a mixed pattern with unilateral obstruction.[15] Cardiac enlargement is usually absent.[13,27]

Diagnosis is confirmed by transthoracic echocardiography (TTE). Preoperative TTE focuses on identifying the pulmonary venous confluence posterior to the left atrium (and lack of communication between the pulmonary confluence and the left atrium), locating the drainage site of each pulmonary vein, identifying any coexisting cardiac abnormalities, and assessing for pulmonary venous obstruction (PVO). Pulmonary venous drainage sites can often be determined without the need for more invasive diagnostic tests.[1,3] Normal pulmonary venous blood flow is low velocity and phasic, varying with the cardiac cycle. Nonphasic venous flow on pulsed Doppler echocardiography is an

indication of obstruction. Flow acceleration proximal to the site of obstruction may also be observed but can be absent in cases of diminished pulmonary blood flow.[1] Computed tomography angiography is helpful in cases where TTE is unable to clearly identify the pulmonary venous drainage sites or the site of obstruction.[27,28]

Cardiac catheterization is generally unnecessary but may be helpful to measure pulmonary artery pressure in children who present later in life or to clarify unusual anatomic variants.[3,8,15] Catheterization is rarely performed in critically ill neonates as it may worsen cardiopulmonary function and delay surgical correction.[11] In fact, avoidance of catheterization is associated with reduced surgical mortality in critically ill neonates.[29] Diagnostic imaging for TAPVC is described in further detail elsewhere in this volume by Files et al.

The optimal timing of surgical correction for neonates without evidence of PVO is unclear. Although avoidance of cardiopulmonary bypass in the neonatal period has theoretical advantages, the rate of prolonged mechanical ventilation appears to be reduced by early surgery. Furthermore, persistent exposure of the left heart chambers to abnormal filling may necessitate a longer period of postoperative support as atrial and ventricular remodeling occur.[30]

Perioperative Anesthetic Management

Neonates with severe PVO are often intubated and mechanically ventilated shortly after birth in an attempt to improve their oxygenation and cardiac output. Inotropic support may be initiated prior to surgery. There is little utility to prolonged preoperative stabilization of critically ill neonates as delayed surgery leads to worsening pulmonary edema and pulmonary hypertension. For newborns presenting in extremis, ventilation should be controlled and acidosis corrected as preparations are made for emergent surgery.[11] Preoperative extracorporeal life support (ECLS) is occasionally necessary in extreme cases when surgery must be delayed in a critically ill patient.

Hemodynamic goals before bypass include maintenance of cardiac output and avoidance of worsening pulmonary edema. Pulmonary vasodilators such as nitric oxide can be detrimental in the setting of obstructed pulmonary venous flow by causing worsening pulmonary edema in the face of fixed obstruction. Standard monitors should be used in addition to arterial and central venous pressure monitoring and cerebral oximetry. In critically ill neonates, transthoracic central lines may be placed by the surgeon prior to separation from bypass in order to limit preoperative delay. If transesophageal echocardiography (TEE) is used, it should be done cautiously as the probe in the mid-esophageal position lies directly behind the pulmonary venous confluence and can cause life-threatening pulmonary venous obstruction. Use of a microsized TEE probe may mitigate this hemodynamic effect. Echo evaluation should be promptly aborted and the probe withdrawn if hemodynamic instability develops during the examination.

Repair is typically performed with deep hypothermic circulatory arrest in neonates. Cardiopulmonary bypass with bicaval cannulation and moderate hypothermia may be used in older children. Repair of supracardiac and infracardiac lesions involves longitudinal incision of the venous confluence and creation of an anastomosis to the left atrium, ligation of connections to the systemic venous system, and closure of the ASD. In cardiac TAPVC, the coronary sinus is unroofed as needed and an ASD patch is carefully placed to baffle pulmonary venous flow to the left atrium. A "sutureless" technique of atriopericardial connection without direct anastamosis to the pulmonary veins or pulmonary venous confluence can be used for primary repair or repair of recurrent pulmonary vein stenosis.[31] Although ligation of the vertical vein is generally part of the repair, a few surgeons prefer to leave the vertical vein open in patients with severe obstruction and small, noncompliant left heart chambers. This minimizes the extent of surgical dissection and duration of cardiopulmonary bypass and may contribute to improved hemodynamic stability in the early postoperative period. However, this approach leaves the patient vulnerable to persistent left to right shunt and subsequent heart failure if the vertical vein does not involute spontaneously over time.[32-34] Surgical techniques for repair of TAPVC are described in greater detail by Shaw et al elsewhere in this volume.

After cardiopulmonary bypass, these neonates require inotropic and vasopressor support similar to other patients undergoing neonatal cardiac surgery. Inodilators such as milrinone are particularly helpful in providing both right and left ventricular afterload reduction as well as improved contractility. Correction of TAPVC imposes an acute volume load on the left ventricle, which may be noncompliant and prone to overdistension with excessive fluid administration. The right ventricle may also struggle to maintain cardiac output in the face of elevated pulmonary pressures.

Pulmonary vasodilation is essential as these patients are at significant risk of pulmonary hypertensive crisis. Endothelial dysfunction induced by ischemia, microemboli, complement activation, and pulmonary leukosequestration during cardiopulmonary bypass further exacerbates neonatal pulmonary vasoreactivity.[35] The degree of pulmonary hypertension is related to the presence of preoperative pulmonary hypertension and the duration of cardiopulmonary bypass. Pulmonary hypertensive crises are associated with increased mortality and should be managed aggressively.[35] Hyperventilation with high inspired oxygen concentration, sedation, and muscle

relaxation helps avoid increases in pulmonary artery pressure (PAP). Modified ultrafiltration after separation from bypass reduces biochemical mediators involved in pulmonary vasoconstriction and may reduce the risk of pulmonary hypertension as well as the duration of mechanical ventilation.[36-38] Inhaled nitric oxide (iNO) can be helpful in managing pulmonary hypertension and contributes to an approximately 25% reduction in mean PAP in high-risk patients.[35,39,40] If a trial of iNO fails to decrease the mean PAP, an anatomic source of obstruction should be considered.[35] In the authors' view, iNO should be initiated prior to separation from cardiopulmonary bypass in all neonates with TAPVC who have preoperative evidence of PVO and should be strongly considered in all TAPVC patients. In some cases, ECLS may be necessary to support the patient through the initial period of postoperative pulmonary hypertension and low cardiac output.[41]

Transesophageal echocardiography may be used to assess the repair and ensure the absence of persistent PVO. However, it is important to note that extrinsic compression of the pulmonary veins by the echo probe can result in inaccurate assessment of the degree of obstruction. In these cases, epicardial echocardiography or postoperative TTE may be required to assess the repair. A thorough examination includes global assessment of function, left ventricular filling, wall motion abnormalities, and intracardiac air. Supraventricular arrhythmias occur in approximately 15% of patients but typically revert to sinus within days of the operation. Ventricular arrhythmias are uncommon.[42]

Particularly in TAPVC with PVO, the lungs are edematous with dilated lymphatic channels and thick alveolar walls.[13] Thus, several days of postoperative mechanical ventilation should be anticipated. The mean duration of mechanical ventilation is 6 days for infants with pulmonary hypertensive episodes and 2 days for those without.[11]

Outcomes

Outcomes for isolated TAPVC have improved considerably over the past few decades; mortality has decreased to approximately 10% in the late 1990s to 2000s owing to improved surgical techniques and perioperative care.[4,11,12,43,44] Outcomes of complex TAPVC, however, are still discouraging.[4,5,43,45] In a cohort of 46 patients with a median age of 26 days undergoing surgical repair of TAPVC between 1989 and 2012, overall operative mortality was 19.6%: 6.9% in isolated TAPVC and 41.2% in complex TAPVC.[4] Other authors have found similar mortality rates in recent years.[43,44] Low weight, young age, elevated pulmonary vascular resistance, mixed-type TAPVC, single-ventricle physiology, and aortic cross-clamp time >60 minutes are risk factors for mortality.[4,23,42] In this series, late mortality at a median follow up of 3 years was 24%.[4]

Common postoperative complications include pulmonary hypertensive crisis, low cardiac output, renal failure, and arrhythmia, as well as other standard sequelae of cardiac surgery.[4,11] Most late deaths occur within the first year after surgery.[4,11,43] Survivors are often asymptomatic; however, even asymptomatic survivors frequently have impaired exercise tolerance related to LV diastolic dysfunction.[4,46]

About 15% of patients develop postoperative PVO, typically within 6 months of surgery. Obstruction may be related to anastomotic stricture or to progressive intimal hyperplasia and fibrosis of the pulmonary veins, which is often fatal.[4,11,47] Patients with diffusely small pulmonary veins, major collaterals, or mixed TAPVC are at greatest risk of death from PVO.[47] Interventions for postoperative PVO range from balloon angioplasty to surgical excision of stenotic segments or sutureless repair. Early intervention is advised to avoid progressive and irreversible pulmonary vascular changes.[47]

Summary

Outcomes for TAPVC have improved dramatically over the past few decades, and mortality for isolated TAPVC is less than 10% in some centers. Outcomes for complex TAPVC remain unfavorable. Newborns with obstructed TAPVC typically present with severe cardiovascular decompensation and require urgent surgical management. Perioperative management focuses on providing inotropic support and managing potential pulmonary hypertensive episodes. Postsurgical survivors without recurrent PVO typically have reasonable functional status and long-term survival.

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