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Total Anomalous Pulmonary Venous Connection: Preoperative Anatomy, Physiology, Imaging, and Interventional Management of Postoperative Pulmonary Venous Obstruction

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Abstract

Total anomalous pulmonary venous connection refers to a spectrum of cardiac anomalies where the pulmonary veins fail to return to the left atrium and the pulmonary venous blood returns through a systemic vein or directly to the right atrium. There is a wide anatomical variety of venous connections and degrees of pulmonary venous obstruction that affect the presentation, surgical repair, and outcomes. In this review, we explore the preoperative physiology, echocardiographic diagnosis, and approach to postoperative complications.

Keywords

congenital heart disease, pediatric intensive care, neonate

Introduction

Total anomalous pulmonary venous connection (TAPVC) refers to a spectrum of cardiac anomalies where the pulmonary veins fail to return to the left atrium. The pulmonary veins return either to one of the systemic veins or directly to the right atrium. There is complete mixture of pulmonary and systemic venous return in the right atrium and an obligate right to left atrial shunting to sustain life. There are multiple anatomical subtypes, and the presentation and outcomes are largely influenced by any point of obstruction to the pulmonary venous return to the heart. An understanding of these anatomical subtypes as well as detailed imaging and physiological assessment are critical to medical and surgical management.

Background and Epidemiology

TAPVC is a rare cardiac malformation with a reported incidence of around 7 per 100 000 live births.[1,2] Although it is normally encountered as an isolated defect, TAPVC has been reported with single-gene disorders such as Holt-Oram and Noonan syndromes.[3] The majority of patients with TAPVC have no family history of congenital heart disease, despite case reports of inheritance in a dominant fashion in siblings and first-degree relatives.[3-5] There are no known environmental fetal factors for TAPVC. One case series reports a male predominance of TAPVC to the portal vein (3.6:1) but not for other sites of TAPVC.[6] In patients with heterotaxy syndrome—a malformation syndrome with abnormal left/right sidedness of the thoracic and abdominal organs—TAPVC occurs with complex cardiac malformations and often contributes to the severity and poor prognosis of this disease. The majority of cases of TAPVC, however, are not associated with significant other cardiac malformations other than a patent foramen ovale and patent ductus arteriosus, and this form of *simple TAPVC* forms the rest of this review.

Embryology

Understanding the various subtypes of TAPVC requires an understanding of the development of the normal pulmonary system and its associated vasculature.[7] The primitive lung buds develop as an anterior division of the foregut. At this time, the primitive lung is drained by the vascular plexus of the foregut, the splanchnic plexus, without any direct connection to the heart. As the pulmonary vascular plexus develops further, the drainage is to the umbilicovitelline

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and cardinal veins.[8] The pulmonary vascular bed then connects to the left atrium by the development of a common pulmonary vein, which reaches the wall of the left atrium. Thus, for a brief period, there is dual drainage to the left atrium (via the common pulmonary vein) and the systemic veins (via the left and right cardinal veins and umbilicovitelline veins). In normal development (approximately 3-5 weeks gestation), the vascular connection to the cardinal and umbilicovitelline veins regresses and the common pulmonary vein further incorporates to become the smooth walled portion of the left atrium (located between the left and right horns of the sinus venosus and to the left of the septum primum).[9] TAPVC results from atresia of the common pulmonary vein or failure of the common pulmonary vein to enter into the left atrium. This typically happens when there are persistent connections to the cardinal or umbilicovitelline veins, thus explaining the classic types of TAPVC.[9] Persistent connection to the left cardinal vein results in TAPVC to the coronary sinus or left innominate vein. Persistent connection to the right cardinal vein results in TAVPC to the right superior vena cava or directly to the right atrium. Finally, persistent connection to the umbilicovitelline veins results in infracardiac TAPVC to the portal veins, ductus venosus, or inferior vena cava. Rarely, late atresia of the common pulmonary vein (after the primitive connections have obliterated) results in the pulmonary veins ending in a blind cul-de-sac, which carries a poor prognosis. There are often diminutive connections from the pulmonary veins to the paraverterbral or splanchnic plexuses, which do not serve as effective decompression from pulmonary venous hypertension.

Stenosis of the common pulmonary vein as it enters into the left atrium results in cor triatriatum, which may demonstrate similar physiology to obstructed TAPVC; this will not be discussed further here. Some or all of the pulmonary veins can have anomalous drainage but normal connections between the right and left horns of the sinus venosus. An example of total anomalous pulmonary venous drainage but normal connection occurs with left deviation of the septum primum, such that the pulmonary veins are connected appropriately, but their drainage is into the right atrium. Partial anomalous pulmonary venous connection (PAPVC) occurs when some of the pulmonary veins connect normally to the left atrium but one or more drain anomalously (with the same embryological basis as described above). Because the presentation, natural history, and treatment are different, PAPVC will not be discussed here.

In utero pulmonary venous obstruction (PVO) can lead to pathological histological changes. Autopsy series have demonstrated arterialization and/or hypoplasia of the pulmonary veins as well as pulmonary lymphangectasia in newborns with fetal PVO. Pulmonary lymphangectasia is a condition characterized by pulmonary lymphatic dilation that presents as an isolated congenital disorder or secondary to PVO. A confirmatory diagnosis is made based on lung biopsy and carries a grave prognosis. Patients with pulmonary lymphangectasia secondary to obstructed TAPVC generally have poor survival despite satisfactory surgical relief of PVO.[1]

Anatomical Subtypes

TAPVC is generally classified into 4 anatomical subtypes: supracardiac (to the innominate vein or right superior vena cava), cardiac (to the coronary sinus or directly to the right atrium), infracardiac, or mixed (see figure from the accompanying article, Figure X). A summary of 3 autopsy series representing the anatomical subtypes shows connections to the left innominate vein (33%), superior vena cava (13%), coronary sinus (11%), right atrium (9%), portal system (19%), and mixed connection (7%).[8] Further description of anatomical subtypes is found in the imaging section.

Fetal Physiology

Because of the high pulmonary vascular resistance in utero, pulmonary blood flow is relatively low. The most easily appreciated hemodynamic alteration of TAPVC on the developing fetus is a reduction in the size of the left atrium and occasionally the left ventricle. The left atrial size is small likely secondary to both morphological and hemodynamic effects. Because the common pulmonary vein normally is incorporated in the posterior left atrium, hearts with TAPVC lack this contribution to the size of the left atrium, and thus, the atrial volume is reduced. It is possible that the reduced size of the left atrium decreases compliance, further reducing flow through the foramen ovale, and thus, both the left atrium and left ventricle may be underdeveloped. The right atrium and right ventricle expand and preserve the total cardiac output and may be significantly dilated late in gestation. Whereas the reduced size of the left atrium and ventricle are usually not significant enough to change surgical planning or preclude complete repair, reduced stroke volume is very commonly encountered. Compensatory sinus tachycardia is a very frequent postoperative finding.[10]

Presentation, Physiology, and Natural History

TAPVC remains the critical cardiac disease with the lowest prenatal detection rate. Routine ultrasound screening in midgestation detects TAPVC in only 2% to 10% of cases.[11] Recent pulse oximeter screening before hospital discharge has picked up asymptomatic children with mild hypoxemia. The clinical presentation, natural history, and outcomes are largely dependent on the anatomical subtype

and the presence of PVO. Although asymptomatic patients without PVO have been reported well into adulthood, the vast majority will become symptomatic in early childhood and are diagnosed within the first year of life.

TAPVC Without PVO

When there is free egress of pulmonary blood flow through the anomalous pulmonary connection back to the right atrium and subsequently across the foramen ovale, there are usually minimal symptoms at birth, and cyanosis may be minimal. All the pulmonary and systemic blood returns to the right atrium, creating a complete admixture. As pulmonary vascular resistance falls in the first several days of life, massive pulmonary overcirculation develops, and increased volume and pressure is transmitted to the right atrium and right ventricle. The cardiac output is maintained by right to left atrial shunting. The degree of cyanosis is variable, but oxygen saturations can be as high as the mid-90s, which is explained by a >5:1 pulmonary to systemic shunt. Although symptoms are minimal at birth, tachypnea and feeding difficulties manifest, followed by profound failure to thrive and death before a year in the majority.[12] Patients demonstrate tachypnea, tachycardia, and hepatomegaly. Cardiac exam either has no murmur or a soft ejection-type pulmonary murmur and, commonly, a S3 gallop. Electrocardiography reveals right atrial and right ventricular enlargement and occasionally an incomplete right bundle pattern. Echocardiography is described below.

TAPVC With PVO

PVO is found in 25% to 50% of TAPVC patients[13-15] and dramatically alters the presentation, physiology, and outcomes. The degree of obstruction exists on a spectrum, ranging from mildly increased pulmonary venous pressure to pulmonary vein atresia associated with catastrophic respiratory failure. Pulmonary venous hypertension and edema progress in the first several hours after birth as pulmonary blood flow increases. Pulmonary edema causes reflexive pulmonary artery vasoconstriction and impaired gas exchange, resulting in progressive hypoxemia. The symptoms and chest radiographic findings can sometimes be difficult to distinguish from neonatal respiratory distress syndrome; obstructed TAPVC may be minimally symptomatic in the first hours after birth but then tends to have a progressive course of hypoxemia and acidosis despite aggressive management. The degree of hypoxia is variable but is often severe and poorly responsive to supplemental oxygen in cases of severely obstructed TAPVC.

The physical findings of obstructed TAPVC often include severe respiratory distress, yet subtle cardiac findings.[8] The precordium often does not have a murmur, but tachycardia and hepatomegaly are typically present. Prompt echocardiography is required to quickly differentiate other forms of neonatal respiratory distress. Prostaglandin E1 (PGE) is often instituted for refractory neonatal respiratory distress with presumed ductal-dependent congenital heart disease. Occasionally, infants worsen with institution of PGE, presumably as a result of increases in the left to right shunt at the ductus arteriosus exacerbating pulmonary venous congestion and edema.

Imaging

Echocardiography provides complete anatomical details to allow for surgical planning in the majority of cases. In general, echocardiographic evaluation of pulmonary veins should include 2D documentation of the number, size, and connection of each pulmonary vein as well as the position of the atrial septum in relation to the pulmonary veins. Color and spectral Doppler are extremely important in locating the pulmonary veins, determining the course, and assessing evidence of obstruction.

Some features are common to all forms of TAPVC. There is significant right atrial and right ventricular volume overload. The left atrium is frequently small and the left ventricle compressed by the dilated right ventricle.[8] There is an obligatory right to left shunt at the atrial level and, frequently, an echo-free space posterior to the left atrium, which represents the pulmonary venous confluence. The entire course of each pulmonary vein and its connection to the central pulmonary venous confluence should be documented by 2D and color Doppler to rule out separate pulmonary vein drainage (mixed type). Additionally, the size and relationship (horizontal vs vertical) of the pulmonary vein confluence to the back of the left atrium aids in guiding the surgical approach.[16] Determining the presence or absence of restriction of flow at the atrial septum is critical. Assessment of right ventricular volume, pressure, and function aids in determining surgical urgency. Frequently, the ductus arteriosus will remain open and allows a precise understanding of the pulmonary pressures. In cases of restricted left heart filling, the flow from the PDA produces retrograde filling of the aortic arch, reflecting poor left ventricular stroke volume. Finally, a complete anatomical study is required to fully evaluate any associated defects.

Often a child will present with extreme respiratory failure, and an urgent but brief echocardiographic assessment is required to aid in treatment. This aids in the rapid planning needed to treat different etiologies of severe respiratory failure (ie, extracorporeal support vs cardiac surgery). In these cases, select views can aid in the rapid differentiation of TAPVC with obstruction from other forms of respiratory failure. Frequently, pulmonary edema makes acoustic windows poor, and thus, subxiphoid images are superior to parasternal ones. Rapid assessment of the atrial

septum from the subxiphoid short and long axes can evaluate for the presence of right to left shunting. An inability to view the pulmonary veins entering the left atrium from these views should prompt a high suspicion for TAPVC. Frequently, an echo-free space with continuous or poorly phasic Doppler will lead to a presumptive diagnosis of obstructed TAPVC, and more thorough echocardiographic evaluation can follow.

Color and Spectral Color Doppler Assessment

Color Doppler is invaluable in locating the site and direction of pulmonary vein flow when visualization of the individual pulmonary veins by 2D is incomplete. Lowering the Doppler scale will aid in visualization because of low-velocity flows in the pulmonary veins. Assessment of pulmonary vein obstruction is, therefore, a combination of color Doppler as well as pulsed and continuous wave spectral Doppler.[17] A normal pulmonary venous Doppler pattern shows normal phases of pulmonary venous flow (Figure 1A). This pattern can be perturbed with pulmonary veins, which are unobstructed but demonstrate a high-flow state, such as with a patent ductus arteriosus or ventricular septal defect (Figure 1B). Pulmonary veins with downstream obstruction often have low Doppler amplitude, which may initially be confusing as a normal signal; however, they lack the normal cardiac phasicity (Figure 1C). When this pattern is encountered, a thorough search for any further area of obstruction is required. In an area of discrete narrowing within the pulmonary veins, the color Doppler becomes very turbulent, and spectral Doppler shows high velocity and an elevated mean gradient (Figure 1D).

Supracardiac TAPVC

Parasternal and suprasternal notch windows are ideal to tracing the course of the ascending vertical vein in supracardiac TAPVC.[8] The size and orientation of the pulmonary vein confluence is often best seen from subxiphoid and parasternal windows, and the confluence is frequently horizontal in supracardiac TAPVC. Parasternal windows allow assessment of each pulmonary vein, the orientation of the confluence to the left atrium, and the origin of the vertical vein as it courses cephalad. The vertical vein typically begins leftward and courses anterior to the left pulmonary artery and left mainstem bronchus. It continues cephalad, anterior to the aorta, before joining the innominate vein. Although supracardiac TAPVC is typically unobstructed, 2 important sites of constriction are possible. The connection with the innominate vein can be narrow and cause various degrees of obstruction (Figure 2). Occasionally, the vertical vein passes between the left pulmonary artery and bronchus, which represents an external source of constriction. The innominate vein and SVC are dilated because of increased flow.

The vertical vein can also connect directly to the right sided superior vena cava or to the azygous vein immediately before connecting to the right superior vena cava. The vertical vein can be obstructed if it passes between the right pulmonary artery and the trachea or right mainstem bronchus.

Cardiac TAPVC

Parasternal and subxiphoid windows are ideally suited to identify drainage to the coronary sinus. The coronary sinus is seen as a severely dilated structure posterior to the left atrium. Parasternal views are used to identify all pulmonary veins returning to the coronary sinus, which serves as the confluence. Subxiphoid windows demonstrate the very dilated coronary sinus ostia entering into the right atrium (Figure 3). Rarely, the ostia of the coronary sinus will be an important source of PVO.[8] Pulmonary venous connection directly to the right atrium occurs with a high association with heterotaxy syndrome and, thus, other complex malformations of the heart. The pulmonary veins can connect directly with the right atrium or via a common pulmonary vein or confluence, which empties into the right atrium. Apical and parasternal imaging is important to document individual pulmonary veins and to rule out mixed forms of TAPVC. PVO is uncommon in cardiac TAPVC.

Infracardiac TAPVC

In hearts with infracardiac TAPVC, the vertical vein descends through the esophageal hiatus, just anterior to the esophagus. Subxiphoid and parasternal imaging is optimally suited to delineate drainage, areas of obstruction, and the size and orientation of the pulmonary venous confluence as well as individual pulmonary veins (Figure 4).[8] The vertical vein most commonly drains to the portal vein but can also connect with the ductus venosus, hepatic vein, or directly into the infradiaphragmatic inferior vena cava. Frequently, the pulmonary vein confluence has a vertical orientation with the upper- and lower-pulmonary veins joining separately along the line of the vertical vein, in a pattern that is said to represent an upside down Christmas tree (Figure 5).

Atrial Septum

The atrial septum must be assessed to demonstrate the size and adequacy of the atrial communication because all the systemic cardiac output must progress through the atrial septum. Subxiphoid short- and long-axis views are ideally

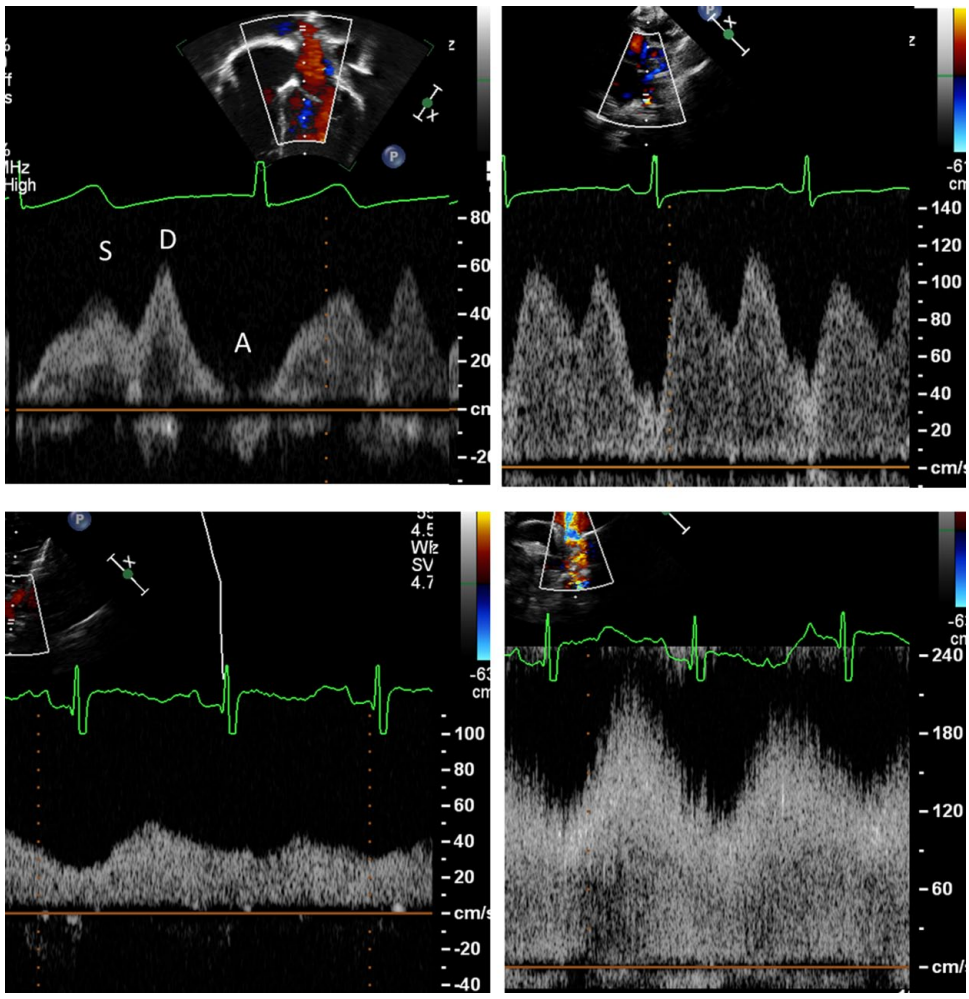

Figure 1. Pulmonary venous spectral Doppler patterns from transthoracic imaging. A. Normal pulmonary vein Doppler from the right upper-pulmonary vein from apical views. The distinct phases of pulmonary venous flow are seen. Flow reversal is typical in atrial systole. B. Unobstructed pulmonary venous pattern with a high-flow state, taken from high parasternal short-axis views. Neonates with high pulmonary blood flow will frequently have elevated velocities relative to normal, but individual pulmonary waveforms are noted. The a-wave returns near baseline during atrial systole for this neonate with truncus arteriosus. C. Pulse-wave Doppler in the left lower-pulmonary vein demonstrates decreased cardiac cycle phasicity and lack of return to baseline or a-wave reversal in this high parasternal short-axis view. This pattern can be seen with mildly obstructed pulmonary veins or pulmonary veins proximal to the site of obstruction. D. Pulse wave Doppler at the connection of the vertical vein to the innominate (from suprasternal notch views) shows high velocity and turbulence consistent with obstruction. The measured mean gradient is 8 mm Hg.
Abbreviations: S, systole; D, early diastolic filling; A, atrial systole.

suited for determining any restriction and the size of the communication.

Additional Imaging Modalities

Echocardiography is the main diagnostic tool for the majority of infants presenting with TAPVC. In rare circumstances, additional imaging is needed to determine the location of pulmonary venous drainage or to identify individual pulmonary veins in forms of mixed TAPVC. Additional imaging is performed if echocardiographic images are not sufficient for surgical planning. The choice between imaging modalities (angiography via cardiac catheterization, computed tomography [CT], or magnetic resonance imaging) depends on each center and the indication.[18,19] At our institution, CT is primarily used when additional imaging is needed because of the high spatial resolution, lack of sedation required, and the short duration of the scan, which are all important features for a critically ill infant.

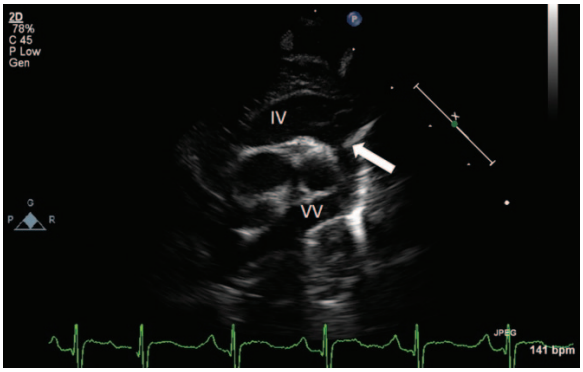

Figure 2. Supracardiac TAPVC to the innominate vein from a transthoracic suprasternal notch view: The vertical vein courses cephalad before entering the innominate vein just medial to the entry of the jugular veins. Minor 2D narrowing (arrow) is noted as the vessel enters the innominate vein. The innominate vein is severely dilated. The spectral Doppler from this patient is found in Figure 1D.
Abbreviations: TAPVC, total anomalous pulmonary venous connection; IV, innominate vein; VV, vertical vein.

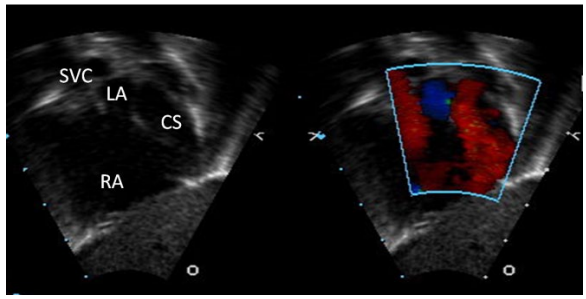

Figure 3. Transthoracic echocardiogram with subxiphoid short axis of TAPVC to the coronary sinus: the severely enlarged coronary sinus returns to the right atrium. Additionally, the right to left atrial shunt is visualized (blue color Doppler).
Abbreviations: TAPVC, total anomalous pulmonary venous connection; SVC, superior vena cava; LA, left atrium; CS, coronary sinus; RA, right atrium.

Surgical Timing

In general, surgical repair is indicated at the time of diagnosis. For infants with respiratory failure and PVO, this remains one of the rare causes of emergent cardiac surgical repair because medical management is often ineffective in stabilizing this inherently unstable physiology. For most term children with TAPVC without obstruction, repair is typically performed within the week after diagnosis. For infants born prematurely or with low birthweight and no evidence of PVO, it may be reasonable to delay surgery to allow for additional somatic growth assuming that children can be watched carefully for symptoms of pulmonary overcirculation and failure to thrive.

Intraoperative Evaluation

Intraoperative transesophageal echocardiography (TEE) is commonplace for assessing preoperative and immediate postoperative anatomy and physiology in most cardiac centers. Special care is taken in the case of TEE for TAPVC. The pulmonary vein confluence is often directly anterior to the esophagus, and the probe can cause compression of the pulmonary vein confluence both preoperatively and postoperatively, especially in neonates weighing less than 3 kg. If TEE is used for this indication, care must be taken to evaluate for clinical signs of hemodynamic instability, desaturation, and respiratory insufficiency. Epicardial imaging can be used to check the adequacy of the pulmonary venous anastomosis if there is concern for probe compression of the surgical repair. Postoperative imaging should also detail right and left ventricular function, atrioventricular valve function, and assessment of right ventricular systolic pressures.

Interventional Management

Transcatheter options exist for both the diagnosis and initial palliation of TAPVC as well as management of resultant PVO following surgical repair. Initial diagnosis is typically made with echocardiography. However, under certain circumstances in which the course of all the pulmonary veins cannot be clearly delineated by ultrasound, a diagnostic catheterization can provide detailed anatomical information prior to surgical repair. In a multi-institutional study of TAPVC management, approximately 10% of patients underwent diagnostic catheterization prior to repair.[1] This approach was more common in mixed-type TAPVC.

Patients presenting with TAPVC are typically managed with primary surgical repair. However, in certain circumstances, a primary transcatheter palliation may be considered. This typically occurs with obstructed TAPVC in the setting of other comorbid conditions that preclude primary surgical repair, such as prematurity, low birth weight, multisystem organ dysfunction, or multiple congenital anomalies. There are a number of case reports and small case series documenting a variety of approaches depending on the subtype of TAPVC.[20-24] Typically, placement of a stent is indicated to relieve obstruction to pulmonary venous return, reduce pulmonary venous hypertension, and allow for stabilization of the patient prior to surgical repair. Vascular access and approach are typically dictated by the anomalous drainage pattern of the pulmonary veins. It is important to have a good

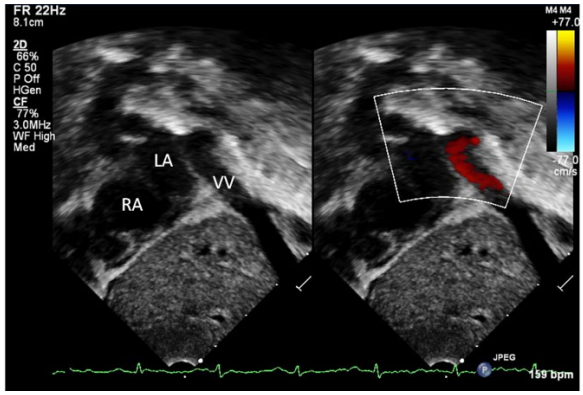

Figure 4. Transthoracic echocardiogram from the subxiphoid short-axis view showing the dilated vertical vein coursing caudally through the diaphragm and into the liver parenchyma. Color Doppler shows caudal direction of pulmonary venous flow toward the diaphragm.
Abbreviations: VV, vertical vein; LA, left atrium; RA, right atrium.

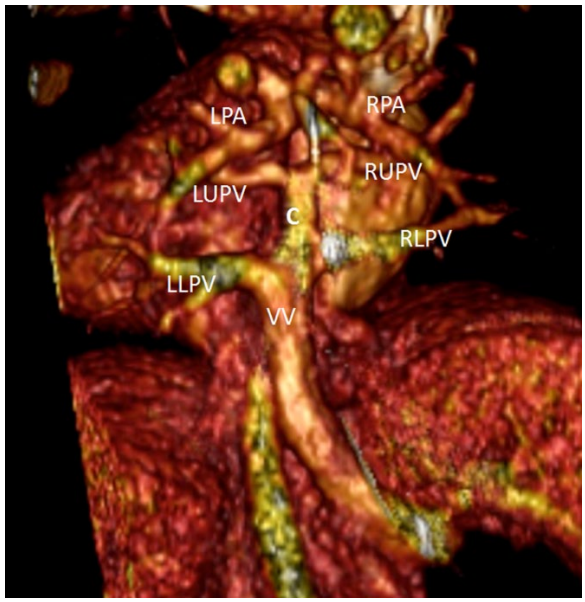

Figure 5. Volume-rendered contrast-enhanced CT (posterior view) of a neonate with infradiaphragmatic TAPVC to the portal venous system. Infradiaphragmatic TAPVC frequently has a vertically oriented confluence (C) with separate entrances of the upper- and lower-pulmonary veins. The vertical vein (VV) is seen as a dilated venous structure descending below the diaphragm and entering the liver parenchyma.
Abbreviations: CT, computed tomography; TAPVC, total anomalous pulmonary venous connection; LPA, left pulmonary artery; RPA, right pulmonary artery; LUPV, left upper-pulmonary vein; RUPV, right upper pulmonary vein; LLPV, left lower-pulmonary vein; RLPV, right lower-pulmonary vein.

understanding of the drainage pattern from preprocedure imaging when planning the intervention.

The majority of transcatheter techniques in TAPVC are directed toward the management of PVO following surgical repair of TAPVC. Reports of the incidence of postoperative PVO vary from 5% to 18%.[1,14,25] The presence of preoperative pulmonary vein hypoplasia or stenosis is a strong risk factor for the development of postoperative disease. Additionally, a single-center study comparing patients with TAPVC and heterotaxy syndrome with those patients without heterotaxy syndrome suggests that need for pulmonary vein reintervention following initial repair is higher in the heterotaxy population.[26] The mechanism of obstruction is believed to be neointimal proliferation, intrinsic pulmonary vein hypoplasia, or inadequate surgical relief of obstruction. Histological studies of primary pulmonary vein stenosis in the absence of anomalous connections suggest that myofibroblast proliferation is responsible for the obstructive lesions.[27,28] Often, PVO in this context can be rapidly progressive, with the majority of patients presenting within the first 6 to 12 months following surgical repair of TAPVC.[29] A number of catheter-based strategies, including conventional and cutting balloon angioplasty as well as drug eluting and bare metal stent (BMS) placement have been described. Studies of conventional balloon angioplasty and cutting balloon angioplasty have demonstrated acute improvements in pulmonary vein gradients as well as vessel luminal diameter.[30,31] However, there is frequently rapid recurrence of significant stenosis requiring multiple repeat interventions. Peng et al[31] report freedom from reintervention at 1 year following cutting balloon angioplasty of 4% compared with 34% for conventional balloon angioplasty. The need for reintervention also portends poor overall survival.

Stent implantation for relief of PVO has also been studied. BMSs, drug-eluting stents (DESs), and covered stents have all been used in this context. In one of the largest single-center studies from Boston Children's Hospital, a total of 47 patients with pulmonary vein stenosis underwent pulmonary vein stent implantation, of which only 9 patients (19%) were included following repair of TAPVC.[32] Stent implantation resulted in acute improvements in right ventricular pressure as well as luminal diameter. The development of in-stent stenosis or neointimal proliferation within the stent was common and rapid. Freedom from diagnosis of significant in-stent stenosis was 37% at 1 year. Luminal diameter >7 mm following stent placement was associated with longer freedom from in-stent obstruction or occlusion. The use of DES in this study was not associated with improved freedom from restenosis or reintervention.

Transcatheter approaches for PVO are frequently performed in patients with significant comorbidities and are associated with risk. Studies of balloon angioplasty have identified adverse events in approximately 25% of cases, including systemic thromboembolic events in 2.7% of cases.[33] The most commonly reported events were catheter-mediated supraventricular tachycardia, pulmonary vein tear or aneurysm, reperfusion edema or hemorrhage, and transient complete heart block. Stent embolization has also been reported in a small number of cases.[32] Despite technical advances, reintervention and mortality remain high for patients with PVO following repair of TAPVC.

Conclusion

TAPVC is a rare cardiac malformation with a wide spectrum of anatomical and hemodynamic variability. Detailed preoperative imaging is critical to ensuring adequate surgical planning and preoperative decision making. For the neonate presenting with severe respiratory failure, prompt anatomical diagnosis of TAPVC with PVO by echocardiography is a critical step for any congenital cardiac center. Although the majority of children with TAPVC will have successful surgical repair and few lifelong sequelae, postoperative PVO remains a difficult problem for the surgeon and interventional cardiologist despite technological advances.

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