

Anomalous Pulmonary Venous Return

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A one-hour-old male is severely tachypneic. He is the product of a full-term spontaneous vaginal delivery to a healthy 27-year-old G1 now P1 mother with good prenatal care. In the delivery room, Apgar scores were 7 at one and five minutes. Central and peripheral cyanosis were present and did not improve with supplemental oxygen.

DIAGNOSIS

What Are the Important Diagnoses to Consider?

Differential diagnosis for this patient is broad and includes:

- transient tachypnea of the newborn;
- meconium aspiration;
- pulmonary hypertension;
- congenital heart disease.

Life-threatening diagnoses such as total anomalous pulmonary venous return (TAPVR) should be ruled out prior to evaluation for less severe disorders.

Is Routine Newborn Pulse Oximetry Screening Useful in Identifying TAPVR?

Not necessarily. Neonates with TAPVR may not be cyanotic at birth. However, pulse oximetry will detect even suboptimal oxygen saturation without frank or severe cyanosis.

Screening pulse oximetry is performed to detect infants with potentially life-threatening duct-dependent lesions or lesions requiring an invasive procedure within the first 28 days of life.

What Clinical Signs and Symptoms are Present in Children with TAPVR?

The signs and symptoms depend of the presence or absence of pulmonary venous obstruction. In patients with obstruction they may include:

- Soft murmur over the vertical vein;
- Cool to cold extremities;
- Cyanosis;
- Tachypnea;
- Hepatomegaly (infracardiac location of anomalous veins);
- Hypotension.

Describe the Anatomy of Pulmonary Venous Drainage in TAPVR

Total anomalous pulmonary venous return is a rare condition in which all pulmonary venous blood empties into the systemic circulation or directly into the right atrium. A right-to-left shunt at the atrial level maintains systemic cardiac output. This may be in the form of a patent foramen ovale (PFO) or atrial septal defect (ASD).

There are four anatomic variants of TAPVR and these are classified according to their relationship with the systemic veins.

Supracardiac TAPVR. The pulmonary veins retain their connection to the embryologic cardinal veins. Pulmonary veins from the left and right lung coalesce and continue as a vertical vein that drains into the right superior vena cava (SVC), azygous vein, or the left SVC. Supracardiac location is the most common variant and is present in nearly 50% of patients with TAPVR (Figure 66.1).

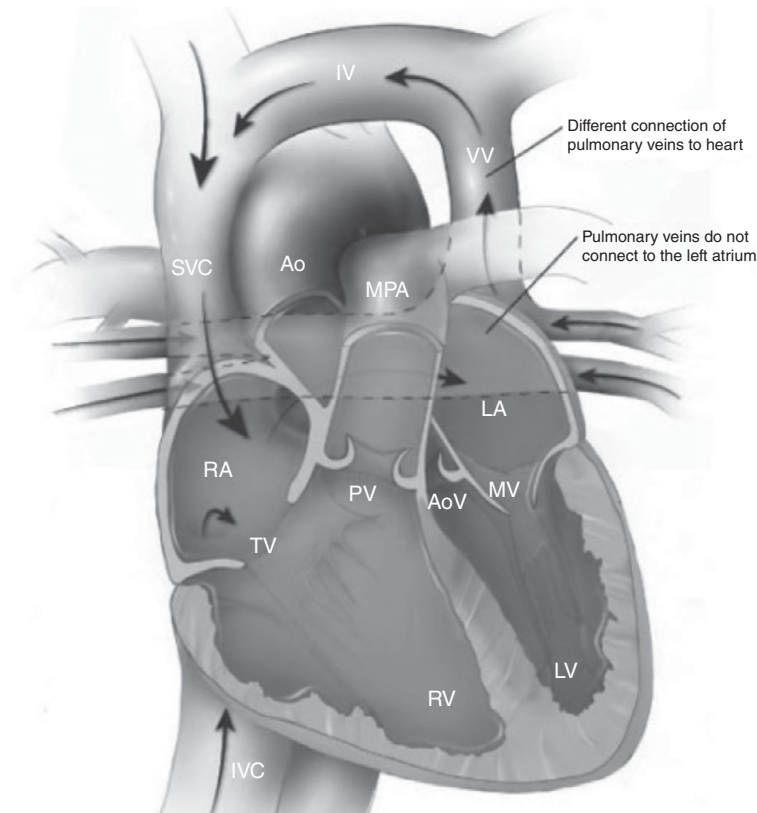


Figure 66.1 Drawing of supracardiac anomalous venous return. The pulmonary veins coalesce posterior to the heart and travel cranially via the vertical vein, eventually draining into the right atrium: IVC, inferior vena cava; SVC, superior vena cava; RA, right atrium; TV, tricuspid valve; RV, right ventricle; PV, pulmonary vein; MPA, main pulmonary artery; LA, left atrium; AoV, aortic valve; LV, left ventricle; MV, mitral valve; PVs, pulmonary veins; VV, vertical vein; IV, innominate vein. Image courtesy of the Centers for Disease Control and Prevention, USA

Cardiac TAPVR. The PVs retain their connection to the embryologic cardinal veins. The veins drain into the right heart directly via the right atrium or via the coronary sinus.

Infracardiac TAPVR. The pulmonary veins (PV) retain their connection to the embryologic umbilicovitelline veins. Pulmonary veins from the left and right lung coalesce and continue as a vertical vein that travels inferiorly through the mediastinum, through the diaphragm, and inserts into the portal vein or inferior vena cava (IVC) below the diaphragm. Due to their lengthy course, infracardiac TAPVR has the highest risk of becoming obstructed.

Mixed TAPVR. Sites of pulmonary venous drainage in mixed TAPVR are variable as the name suggests. Commonly the left and right pulmonary veins have separate drainage patterns. These drainage patterns may involve supra-cardiac, cardiac, or infracardiac locations.

What Anatomic Anomalies Are Associated with TAPVR?

Atrial septal defects (ASD) are nearly universally present, as they allow pulmonary venous blood to enter the left atrium and maintain systemic cardiac output. If a paucity of blood has entered the left-sided heart structures in utero, left-sided heart structures may be small in size. Other associated lesions include: patent ductus arteriosus, ventricular septal defect, and pulmonary stenosis.

Heterotaxy with poly- or asplenia is also common in patients with TAPVR.

What Are the Clinical Presentations of TAPVR?

Clinical presentation is patient dependent and based on the anatomic variant and presence or absence of obstruction. A neonate with obstruction of any of the anatomic variants of TAPVR will be critically ill, severely cyanotic, and have severe pulmonary edema.

Supracardiac TAPVR. Clinical presentation is usually with tachypnea alone as these veins are less commonly unobstructed. An ASD is usually present and allows right-to-left shunting (and left heart filling). The patients are often not markedly cyanotic as the larger shunt from the lesion is left to right (pulmonary veins to right atrium).

Cardiac TAPVR. As pulmonary veins of cardiac TAPVR are least likely to be obstructed, presentation may be subtle. Saturated blood from the pulmonary veins usually drains into the left atrium via an unroofed coronary sinus (CS) or ASD.

Infracardiac TAPVR. Pulmonary veins of infracardiac TAPVR are very likely to be obstructed. Clinical presentation is often of a tachypneic, cyanotic infant in significant respiratory distress with pulmonary edema.

Mixed TAPVR. As expected, presentation is usually dictated by degree of obstruction of pulmonary venous drainage.

How Is the Diagnosis of TAPVR Established?

Pre- or postnatal echo can suggest the diagnosis. Transesophageal echocardiography is not performed as placement of the echo probe may compress any pulmonary venous confluence posterior to the left atrium and result in rapid cardiovascular collapse.

How Is Pulmonary Venous Obstruction (PVO) Defined?

Suboptimal oxygen saturation in addition to nonphasic flow velocity (on echo) greater than 1.8 m/s define PVO.

What Are the Physiologic Consequences of PVO?

Veins that are obstructed have consequent pulmonary venous hypertension. This increases pulmonary

capillary pressure and very quickly leads to pulmonary edema. At this point, chest roentgenogram reveals bilateral infiltrates and may be confused with intrinsic lung disease. After the development of pulmonary edema, pulmonary arteries constrict to prevent further edema. Subsequently, right ventricular (RV) pressures increase, and systemic hypoxia ensues.

Hypoxic pulmonary vasoconstriction further increases pulmonary vascular resistance (PVR), exacerbates RV hypertension, and worsens physiologic trespass.

What Is the Natural History of TAPVR?

The clinical course of TAPVR depends on the presence and degree of obstruction.

Patients with severe obstruction require emergent surgical intervention.

Patients with restrictive intra-atrial communication (small ASD) require intervention to enlarge this communication and maintain cardiac output and oxygenation.

For those who survive infancy, repair of TAPVR has a good prognosis (Reddy et al., 2011). In each of the 26 patients reported, there were no early deaths or postoperative complications. None in this series had infracardiac TAPVR. Of those with significant RV dysfunction, a 5 mm ASD was left open postoperatively. Factors attributed to survival beyond infancy include a large ASD and near normal pulmonary vascular resistance.

Patients with unobstructed veins may go unrecognized with mild symptoms. These patients generally suffer from chronic right heart failure due to overcirculation and may develop pulmonary hypertension.

What Is the Short-Term Outcome of Infants Born with TAPVR?

Postoperative survival is reduced in neonates by the presence of pulmonary venous obstruction, mixed TAPVR, single ventricle physiology, or heterotaxy. Early mortality postoperatively occurs in about 5% of patients. Patients with single ventricle anatomy have a high mortality and morbidity due to chronic pulmonary vein stenosis requiring reoperation.

What Is the Long-Term Outcome of Infants Born with TAPVR? What Cardiac Interventions Are often Required Years Following the Initial Repair?

Severe and recurrent pulmonary vein obstruction is a leading cause of late mortality. After correction of TAPVR, pulmonary venous confluence stenosis remains a long-term complication. While percutaneous interventions have been successful in the form of PV stents, chronic obstruction remains a major source of morbidity.

What Are the Specific Anesthetic Concerns Related to Patients with TAPVR?

Patients with TAPVR require management of pulmonary blood flow as they generally have some degree of pulmonary overload. This may result in chronic pulmonary venous congestion and reduced lung compliance. High inspired oxygen concentration is often required in addition to higher levels of positive end-expiratory pressure (PEEP). Avoidance of metabolic acidosis is important and attempts to correct base deficits should be considered when appropriate.

For a variety of reasons (circulatory overload, small left ventricle, associate anomalies) these patients may require inotropic support.

Transesophageal echocardiography is usually avoided as it may worsen obstruction by external compression, especially in neonates.

Pre-bypass, inhaled nitric oxide is avoided as this may increase pulmonary blood flow and worsen

cyanosis. Post-bypass, inhaled nitric oxide is often used and should be immediately available to reduce pulmonary vascular resistance.

Post-bypass, the left heart, which is often small, may be ill-prepared to accept the full complement of pulmonary venous return.

In patients post repair of anomalous veins, recurrent venous obstruction should be considered.

How Is TAPVR Different from PAPVR?

PAPVR or partial anomalous pulmonary venous return is where one or more (but not all) of the pulmonary veins drains outside the left atrium. Most commonly a single vein drains into the SVC and is a source of persistent left-to-right shunt. Chronic right heart overload can, over time, lead to symptoms of congestion however PAPVR, especially single anomalous veins, is often an incidental finding and does not require treatment.

What Are the Specific Anesthetic Concerns Related to Patients with PAPVR?

Patients with one or more anomalous veins (especially supracardiac veins entering the SVC) are considered for a Warden Procedure. This procedure involves removing the portion of the SVC with the anomalous veins connected and baffling this blood to the left atrium. The SVC flow is reestablished with the use of a small conduit. As such, issues related to the baffle as well as the SVC conduit may occur, especially stenosis.

Suggested Reading

DiBardino DJ, McKenzie ED, Heinle JS, et al. The Warden procedure for partially anomalous pulmonary venous connection to the superior caval vein. *Cardiol Young*. 2004;14(1):64–7. PMID: 15237673.

Mahle WT, Newburger JW, Matherne P, et al. Role of pulse oximetry in

examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. *Pediatrics*. 2009;124(2):823–6. PMID: 19581259.

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Cardiovasc Thorac Ann. 2011;19:249–52. PMID: 21885551.

Shi G, Zhu Z, Chen J, et al. Total anomalous pulmonary venous connection: the current management strategies in a pediatric cohort. *Circulation*. 2017;135(1):48–58. PMID: 27881562.