# Pulmonary and Systemic venous anomaly – Morphology, Surgical repair and postoperative management

연세대학교 심장혈관병원 심장혈관외과

박한기

**Venous Anomalies of the Thorax**

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|---|---|---|
| Pulmonary vasculature | Partial anomalous pulmonary venous return | Right lung drainage to<br>- SVC<br>- Right atrium<br>- Coronary sinus<br>- Azygous vein<br>- IVC-Scimitar, Pseudoscimitar syndrome<br>Left lung drainage to<br>- Left innominate vein via vertical vein<br>- Coronary sinus<br>- Hemiazygos vein |
| | Total anomalous pulmonary venous return | Supracardiac<br>Cardiac<br>Infracardiac<br>Mixed |
| Systemic vasculature | Anomalies of SVC | Persistent left SVC<br>- Single left SVC draining into coronary sinus or left atrium<br>- Duplicated connected by brachiocephalic vein<br>- Duplicated not connected by bacheocephalic vein<br>Right SVC<br>- Drainage to left atrium<br>- Anomalous left brachiocephalic vein drainage to right SVC |
| | Anomalies of Azygos vein | Azygos lobe<br>Hemiazygos lobe<br>Absent azygos vein<br>Azygos continuation<br>Hemiazygos continuation drainage to<br>- Left SVC<br>- Right SVC |

## Partial Anomalous Pulmonary Venous Return

**Anatomy**

Right upper pulmonary vein draining to the superior vena cava is the most common type of partial anomalous pulmonary venous return (PAPVR). This type of partial anomalous pulmonary venous return is commonly associated with sinus venosus type ASD. The sinus venous ASDs are located in the posterior atrial septum. Most commonly, one or both right pulmonary veins drain (right upper most commonly) to either the RA or the SVC, just lateral to the ASD. Less commonly, the pulmonary veins may enter the RA close to the IVC-RA junction.

**Surgical Therapy**

Surgical therapy is usually performed through a median sternotomy with bicaval cannulation,

caval snares, and right atriotomy. Sinus venosus type atrial septal defects associated with PAPVR are treated with an intra-atrial patch that baffles the right-sided pulmonary vein flow through the ASD into the LA. Care must be taken to enlarge the ASD, if needed, to avoid obstruction of the baffle. When the anomalous veins drain high into the SVC and the baffle narrows the SVC lumen, the SVC can be patched open or transected above the anomalous pulmonary veins and anastomosed to the RA appendage (Warden procedure). Isolated coronary sinus defects are treated by closing off the coronary sinus.

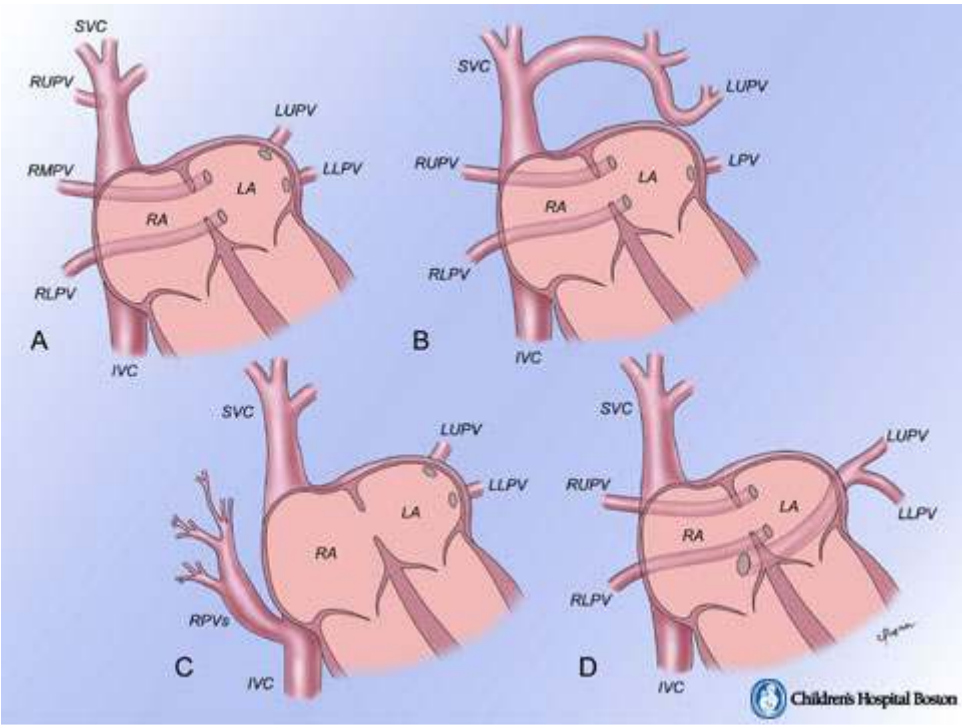


Figure 1. Diagram of partial anomalous pulmonary venous return

## Total Anomalous Pulmonary Venous Return

Total anomalous pulmonary venous return (TAPVR) is a pathologic entity in which all of the pulmonary venous effluent from the lungs drains to the systemic venous system, creating a large left-to-right shunt. An obligatory right-to-left shunt must be present to allow blood to reach the LV and contribute to systemic cardiac output. Most commonly, this occurs at the level of the atrial septum, but may present as a VSD or a PDA. The absence of a right-to-left shunt is incompatible with survival.

**Anatomy**

TAPVR is classified by the site of connection to the systemic venous system into:

- *Supracardiac* (50%). Most common. The ascending vertical vein most often drains into the innominate vein.
- *Cardiac* (20%). The venous confluence drains directly into the right side of the heart, most commonly the coronary sinus.
- *Infracardiac* (25%). The pulmonary vein confluence drains via a descending vertical vein to the portal vein, ductus venosus, or IVC. Infracardiac TAPVR is the most common subset to present with obstruction and occurs at the junction of the vertical vein with the systemic venous drainage.
- *Mixed* (5%). Multiple sites of systemic venous return.

PFO or secundum ASD is always present and these are usually non-restrictive.

All patients with TAPVR exhibit a varying degree of lymphangiectasia and media hypertrophy in both the pulmonary arterial and venous circulation. Varying degrees of pulmonary venous obstruction and congestion are present and occur most commonly in the infracardiac type. In patients with complete obstruction, pulmonary hypertension will be present.

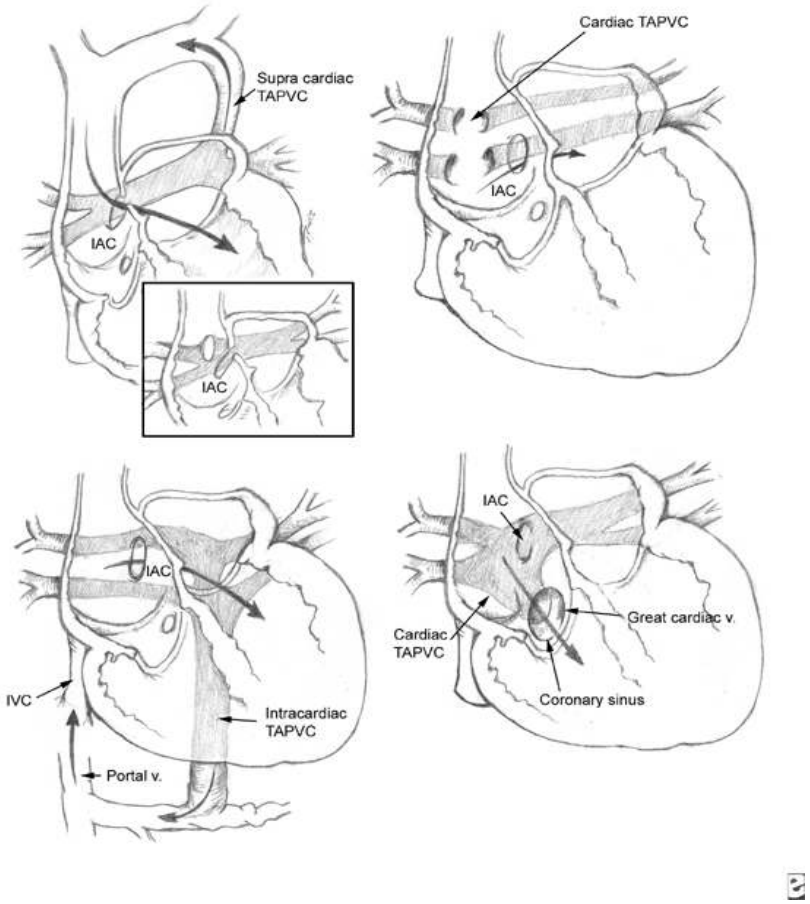


Figure 2. Types of total anomalous pulmonary venous return.

**Presentation**

Patients without significant obstruction present in infancy or childhood with symptoms of heart failure due to their large left-to-right shunt. These patients have dyspnea, poor feeding, and poor growth.

Patients presenting with complete pulmonary venous obstruction constitute a surgical emergency. Expeditious medical measures should be taken to resuscitate the patient, including intubation and mechanical ventilation with 100% oxygen, hyperventilation, correction of acidosis, and inotropic support. Medical measures are minimally effective and surgically correction is required. In patients who present with severe end organ dysfunction, some centers choose a period of ECMO support prior to operative repair. At this time, most centers choose to perform a complete repair with use of ECMO postoperatively only if required.

**Pathophysiology**

The most important anatomic factors in determining the clinical status of the patient include the presence and location of the right-to-left shunt, and the presence or absence of obstruction in the pulmonary venous pathway. As pulmonary venous blood is diverted from the LA, blood is unable to reach the LV in the absence of a right-to-left shunt. In the presence of a shunt, the cardiac output is limited to the amount of blood passing through the right-to-left shunt. A second important anatomic factor that determines the clinical status is the presence or absence of obstruction in the pulmonary venous pathway. With obstruction, egress of blood from the lungs is limited, resulting in pulmonary venous congestion and impairment of oxygenation, which leads to life-threatening neonatal cyanosis. If associated with a restrictive right-to-left shunt and reduced cardiac output, the patient's precarious clinical status is worsened.

An important subset of obstructed TAPVR is patients in whom the vertical vein ascends between the left PA and the left mainstem bronchus. As the degree of pulmonary vein obstruction worsens, PAP increases, causing further distension of the PA and vertical vein compression. Ultimately, this leads to circulatory collapse due to the physiologic vice.

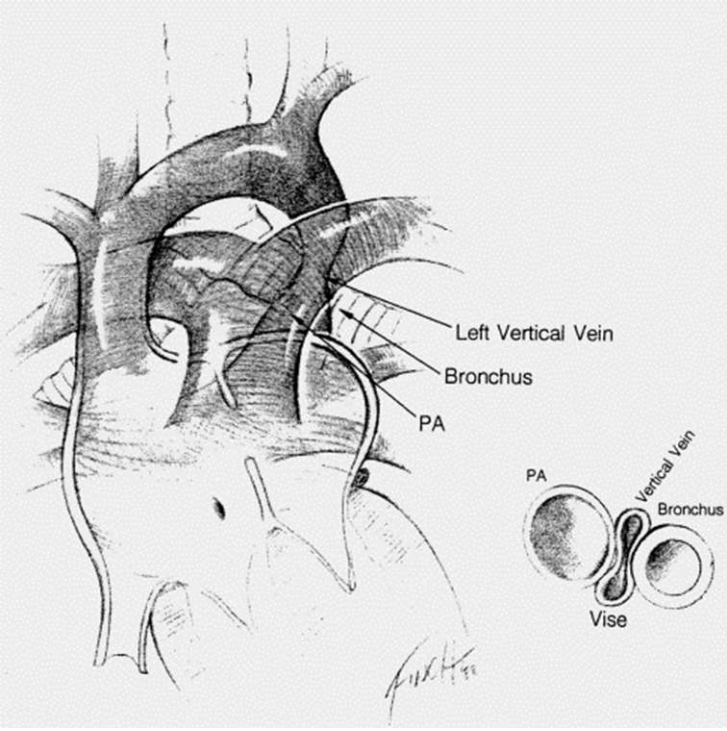


Figure 3. Diagram illustrating compression of the connecting left vertical vein between the left mainstem bronchus and the pulmonary artery

**Diagnostic Techniques**

CXR shows varying degrees of pulmonary venous congestion dependent upon the degree of obstruction. A prominent RA border and PA vasculature may also be seen.

Echocardiography is the study of choice for identifying TAPVR. The pulmonary venous confluence, pulmonary veins, and connection to the systemic venous system can typically be defined. Intraoperative echocardiography is the best modality to evaluate the anastomosis between the venous confluence and the LA. Echocardiography is also the best modality for long-term follow-up.

Cardiac catheterization is infrequently used in a preoperative setting. Indications for angiography include ambiguous anatomy that requires further delineation, or if intervention is required for preoperative stabilization (i.e., balloon atrial septostomy). A classic finding at catheterization is identical oxygen saturations in all chambers of the heart. MRI is emerging as an important diagnostic modality when anatomical clarification is needed and allows a noninvasive method to determine Qp:Qs.

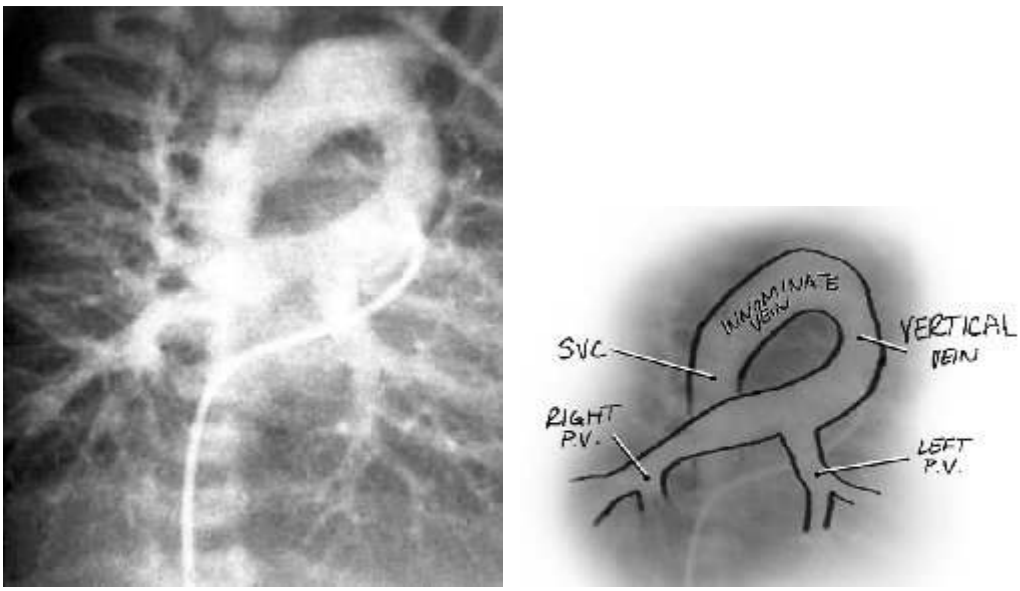


Figure 4. Catheterization of the supracardiac type total anomalous pulmonary venous return

**Surgical Repair**

The treatment of TAPVR is surgical. In asymptomatic patients whose venous outflow is unobstructed by echocardiography, repair is delayed several weeks or until symptoms of dyspnea and feeding intolerance develop. Most children will develop symptoms within 6 months of age. Patients presenting with complete obstruction are considered a surgical emergency, and after attempted medical stabilization, operative repair is performed. The conduct of the operation is indistinguishable when performed emergently or electively. Cannulation is performed with a single arterial and venous. If other intracardiac work is expected, bicaval cannulation is likely required. The patient is cooled to 18 °C and if present, the patent ductus is immediately ligated after initiating CPB. Both supracardiac and infracardiac TAPVR require a direct anastomosis between the pulmonary vein confluence and the LA. With the low-pressure of newborn pulmonary venous blood flow, it cannot be overstated that the anastomosis must be meticulously performed, free of both torsion and tension, to permit unimpeded blood flow. For cardiac variant TAPVR, repair requires unroofing of the coronary sinus into the LA with patch closure of the ASD. In this setting, the drainage from the coronary sinus will be directly into the LA. For patients who have small pulmonary veins or a small venous confluence, a sutureless repair may be required. In a sutureless repair, the anterior wall of the venous confluence is widely opened to the pericardial well. The posterior aspect of the LA is incised and sewn directly to the pericardial well. The pulmonary venous drainage is collected in the newly created pericardial-LA reservoir. The sutureless repair is often the procedure of choice for reoperative pulmonary vein stenosis.

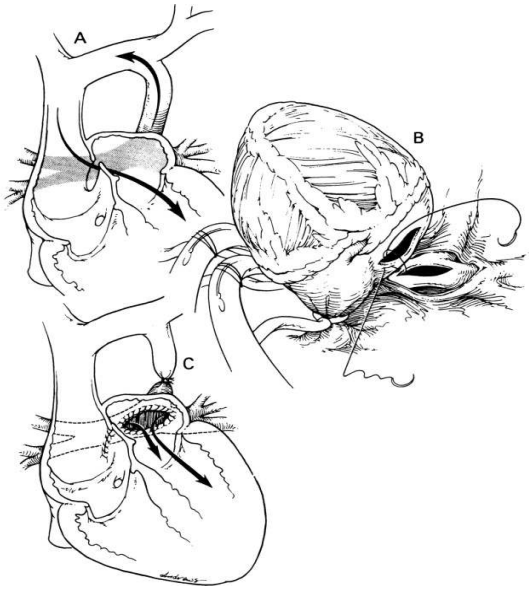


Figure 5. Repair of a supracardiac type total anomalous pulmonary venous connection.

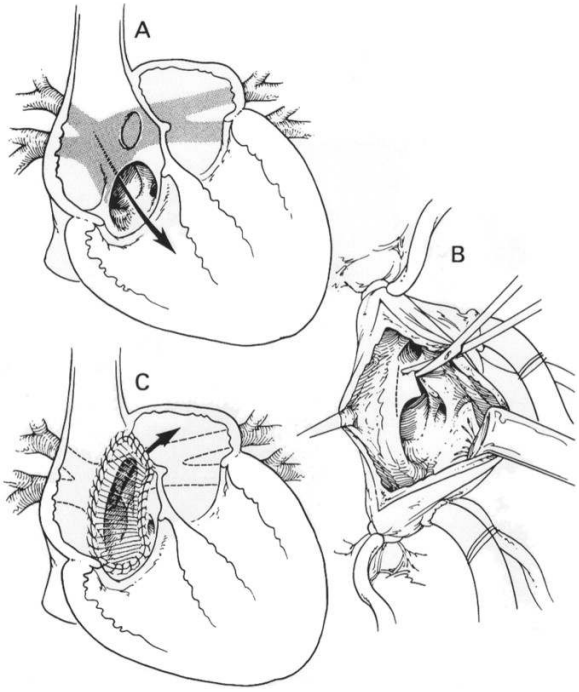


Figure 6. Repair of a total anomalous pulmonary venous connection to the coronary sinus.

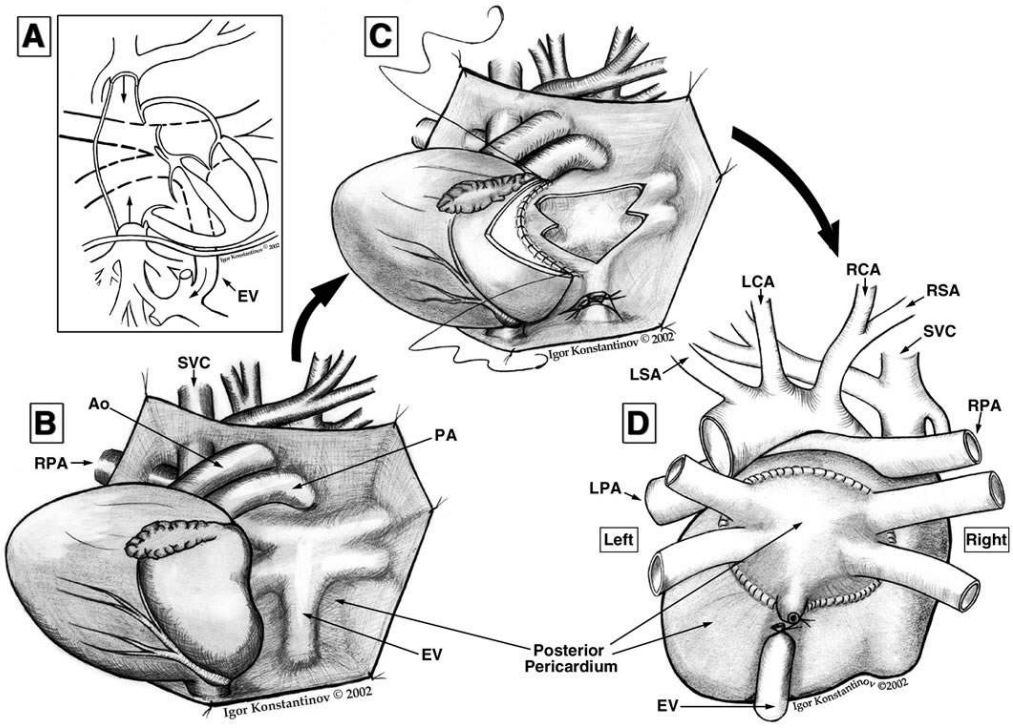


Figure 7. Sutureless pulmonary vein stenosis repair