Pulmonary Venous Anomalies and Obstruction

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**Pulmonary Venous Anomalies and Obstruction**

In patients born with anomalous pulmonary venous connection, pulmonary venous connection and obstruction is a variable factor; thus some patients manifest symptoms of pulmonary venous congestion and congestive heart failure while others do not. It is a relatively common condition of total anomalous pulmonary venous connections to the left innominate vein and to the infradiaphragmatic venous system. In these conditions, the pulmonary veins converge to form a common venous sinus from which an anomalous vertical vein ascends to the innominate vein or descends to join the hepatic portal venous channels, one of its tributaries or the ductus venosus.

Obstruction is reportedly related to "long route" high resistance connections, singly or in combination with crowding of the anomalous pulmonary venous trunk as it passes into the abdomen at the esophageal hiatus (diaphragm) with the esophagus, or the occurrence of localized stenotic lesions in the anomalous channel. Also, the narrowed condition of the ductus venosus or interposition of the hepatic sinusoids between the lungs and the right atrium contributes to the venous obstruction.

**Magnetic Resonance Imaging in Anomalous Pulmonary Venous Connections**

I. **Supracardiac and Cardiac Connections:**

1. **Partial anomalous pulmonary venous return (PAPVR)**

   A congenital process that results from one or more, (but not all) of the pulmonary veins draining into the right atrium. It is typically associated with an atrial septal defect (Sinus venosus or ostium secundum type). Patients are not clinically cyanotic and are asymptomatic. Pulmonary congestion and venous obstruction are rare.
2. Total anomalous pulmonary venous return (TAPVR)

All pulmonary veins are involved with return of pulmonary venous blood flow to the right heart. In this connection an atrial septal defect with left to right shunt is needed for survival. A functional right to left shunt (oxygenated blood) is however present. Cyanosis results as near identical oxygenation develops in all cardiac chambers.

II. Location of Anomalous Connections

A. Supracardiac Form—Most common (Type I)

The anomalous veins drain into an ascending vertical vein that connects with the left brachiocephalic vein and superior vena cava, ultimately emptying into the right heart. This connection is seen in approximately 50% of patients with anomalous pulmonary venous connections.

1. Snowman heart = dilated SVC+ left vertical vein
2. Increased pulmonary vasculature / right heart overload

B. Cardiac Form—(Type II)

The anomalous veins drain directly into a cardiac structure or chamber. It is seen in ~ 30% of patients with anomalous pulmonary venous connections. These veins drain into the coronary sinus or right atrium. The coronary sinus connection occurs most commonly but both lead to right heart volume overload with pulmonary congestion.

C. Infracardiac Form—(Type III)

The anomalous veins in this connection drain below the heart and diaphragm via a descending vertical vein with an elongated venous course. The veins ultimately connect with the inferior vena cava, ductus venosus, portal vein (most commonly) or one of its tributaries. It is seen in approximately 10 to 12% of patients with this form of anomalous venous connection. Venous obstruction typically occurs at the diaphragm - esophageal hiatus. Pulmonary congestion and
cyanosis are resultant outcomes of the admixture of the blood and a right to left atrial shunt. Asplenia (80%) or polysplenia are commonly associated with this anomaly.

D. Mixed Form—(Type IV)

The anomalous veins in this connection drain typically through a supracardiac or cardiac connection as described above. Veins from either the right heart, left heart or both may be involved. This lesion is somewhat rare in this family of lesions. It occurs in 5 to 6% of patients with anomalous pulmonary venous connections.

III. Spin Echo MR Imaging

a. Intracardiac (Intra and Extracardiac connections and lesions)
b. Pulmonary venous connections

IV. Steady State Free precession MR Imaging

a. Anatomy (Intra and Extracardiac connections and lesions)
b. Pulmonary venous connections
c. Venous obstructions

V. Phase Contrast Imaging

a. Venous Flows (Qp:Qs)
b. Venous Gradient Assessment

VI. Magnetic Resonance Angiography and 3-Volume Rendering

VII. Summary