What Is It?

In this defect, the pulmonary veins (PV), which carry oxygen-enriched (red) blood from the lungs to the heart for pumping to the body, are "anomalous," or abnormal with respect to how they return to the heart.

There are various forms of this defect, but all of them involve the red blood from the lungs being carried by the pulmonary veins back to the right side of the heart rather than to the left atrium (LA), as in a normal heart. This means that all the red blood is mixed with the oxygen poor venous (blue) blood in the right atrium. To get to the body, blood must cross through a hole in the atrial septum.

In the most common form of this defect (shown in the illustration), the pulmonary veins (PV) are connected by a vessel to the superior vena cava (SVC), which carries blue (oxygen-depleted) blood back to the heart. This is known as the supracardiac type. There is usually no obstruction with this type of anomalous veins. Less commonly, the pulmonary veins may be connected to the inferior (IVC) vena cava (infracardiac) or to the right atrium itself (intracardiac). It is also possible for the veins to be of mixed type. For example, the left sided veins may return via a supracardiac route and the right-sided veins via an infracardiac route.
What Are Its Effects?

If there is no obstruction to blood flow, infants and children with this defect can be relatively asymptomatic. However, if there is obstruction anywhere along the abnormal course of the blood flow, they can be quite ill.

How Is It Treated?

Usually the pulmonary veins come to a confluence behind the left atrium. Prior to birth, this confluence is supposed to connect to the heart. If it does not, the veins have to find another way to get back to the heart. Surgical treatment of Total Anomalous Pulmonary Venous Return involves reconnecting the pulmonary veins to the back of the left atrium, as in a normal heart.

The illustrations (below) show the repair of the most common type (supracardiac). The left atrium and pulmonary veins are opened and sutured together (1). Notice that the vessel that connects the superior vena cava (SVC) with the pulmonary veins is removed (2). However, this procedure is not always done by the surgeon. It may just be tied off as the blood will bypass this section when it is closed. The atrial septal defect is closed with a patch (3).

The post-operative recovery period depends on the exact nature of the defect and the procedure used to correct it. In those cases where there was an obstruction to the blood flow, corrective surgery may
become urgent and the recovery may be more complicated with a prolonged post-operative course. If the venous blood flow is unobstructed the hospital stay can be as short as 3 to 5 days.

Surgical Repair of Total Anomalous Pulmonary Venous Return