

Description: Rare defect characterized by failure of the pulmonary veins to join the left atrium. Instead, the pulmonary veins are abnormally connected to the systemic venous circuit via the right atrium or various veins draining toward the right atrium, such as the SVC. The abnormal attachment results in mixed blood being returned to the right atrium and shunted from the right to the left through an ASD. TAPVC (also called *total anomalous pulmonary venous return* or *total anomalous pulmonary venous drainage*) is classified according to the pulmonary venous point of attachment as follows:

Supracardiac: Attachment above the diaphragm, such as to the SVC (most common form) (see Fig. 23-10)

Cardiac: Direct attachment to the heart, such as to the right atrium or coronary sinus

Infradiaphragmatic: Attachment below the diaphragm, such as to the IVC (most severe form)

Pathophysiology: The right atrium receives all the blood that normally would flow into the left atrium. As a result, whereas the right side of the heart hypertrophies, the left side, especially the