left atrium, may remain small. An associated ASD or patent foramen ovale allows systemic venous blood to shunt from the higher pressure right atrium to the left atrium and into the left side of the heart. As a result, the oxygen saturation of the blood in both sides of the heart (and ultimately in the systemic arterial circulation) is the same. If the pulmonary blood flow is large, pulmonary venous return is also large, and the amount of saturated blood is relatively high. However, if there is obstruction to pulmonary venous drainage, pulmonary venous return is impeded, pulmonary venous pressure rises, and pulmonary interstitial edema develops and eventually contributes to HF. Infradiaphragmatic TAPVC is often associated with obstruction to pulmonary venous drainage and is a surgical emergency.

Clinical manifestations: Most infants develop cyanosis early in life. The degree of cyanosis is inversely related to the amount of pulmonary blood flow—the more pulmonary blood, the less cyanosis. Children with unobstructed TAPVC may be asymptomatic until pulmonary vascular resistance decreases during infancy, increasing pulmonary blood flow with resulting signs of HF. Cyanosis becomes worse with pulmonary vein obstruction; when obstruction occurs, the infant's condition usually deteriorates rapidly. Without intervention, cardiac failure will progress to death.

Surgical treatment: Corrective repair is performed in early infancy. The surgical approach varies with the anatomic defect. In general, however, the common pulmonary vein is anastomosed to the back of the left atrium, the ASD is closed, and the anomalous pulmonary venous connection is ligated. The cardiac type is most easily repaired; the infradiaphragmatic type carries the highest morbidity and mortality because of the higher incidence of pulmonary vein obstruction. Potential postoperative complications include re-obstruction; bleeding; dysrhythmias, particularly heart block; PAH; and persistent heart failure.

Prognosis: Mortality is between 5% to 10% for infants without obstruction, and it can be as high as 20% for infants with infradiaphragmatic type (Park, 2014).