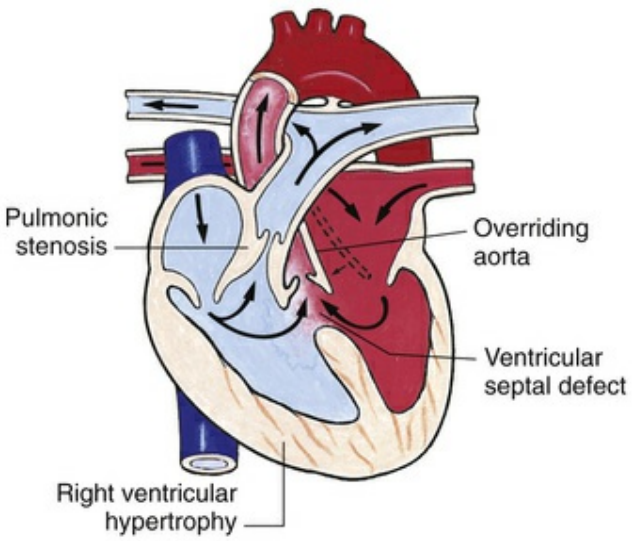

Description: The classic form includes four defects: (1) VSD, (2) PS, (3) overriding aorta, and (4) right ventricular hypertrophy. Tetralogy of Fallot occurs in 5% to 10% of all CHDs and is the most common cyanotic lesion (Park, 2014).

Pathophysiology: The alteration in hemodynamics varies widely, depending primarily on the degree of PS but also on the size of the VSD and the pulmonary and systemic resistance to flow. Because the VSD is usually large, pressures may be equal in the right and left ventricles. Therefore, the shunt direction depends on the difference between pulmonary and systemic vascular resistance. If pulmonary vascular resistance is higher than systemic resistance, the shunt is from right to left. If systemic resistance is higher than pulmonary resistance, the shunt is from left to right. PS decreases blood flow to the lungs and consequently the amount of oxygenated blood that returns to the left side of the heart. Depending on the position of the aorta, blood from both ventricles may be distributed systemically.

Clinical manifestations: Some infants may be acutely cyanotic at birth; others have mild cyanosis that progresses over the first year of life as the PS worsens. There is a characteristic systolic murmur that is often moderate in intensity. There may be acute episodes of cyanosis and hypoxia, called *blue spells* or *tet spells*. Anoxic spells occur when the infant's oxygen requirements