

ventricle and out to the lungs. The condition is often associated with PS and TGA. There is complete mixing of unoxygenated and oxygenated blood in the left side of the heart, which results in systemic desaturation, and varying amounts of pulmonary obstruction, which causes decreased pulmonary blood flow.

Pathophysiology: At birth, the presence of a patent foramen ovale (or other atrial septal opening) is required to permit blood flow across the septum into the left atrium; the PDA allows blood flow to the pulmonary artery into the lungs for oxygenation. A VSD allows a modest amount of blood to enter the right ventricle and pulmonary artery for oxygenation. Pulmonary blood flow usually is diminished.

Clinical manifestations: Cyanosis is usually seen in the newborn period. There may be tachycardia and dyspnea. Older children have signs of chronic hypoxemia with clubbing.

Therapeutic management: For neonates whose pulmonary blood flow depends on the patency of the ductus arteriosus, a continuous infusion of prostaglandin E₁ is started at 0.1 mcg/kg/min until surgical intervention can be arranged.

Surgical treatment: Palliative treatment is the placement of a shunt (pulmonary-to-systemic artery anastomosis) to increase blood flow to the lungs. If the ASD is small, an atrial septostomy is performed during cardiac catheterization. Some children have increased pulmonary blood flow and require pulmonary artery banding to lessen the volume of blood to the lungs. A bidirectional Glenn shunt (cavopulmonary anastomosis) may be performed at 4 to 9 months as a second stage.

Modified Fontan procedure: Systemic venous return is directed to the lungs without a ventricular pump through surgical connections between the right atrium and the pulmonary artery. A fenestration (opening) is sometimes made in the right atrial baffle to relieve pressure. The patient must have