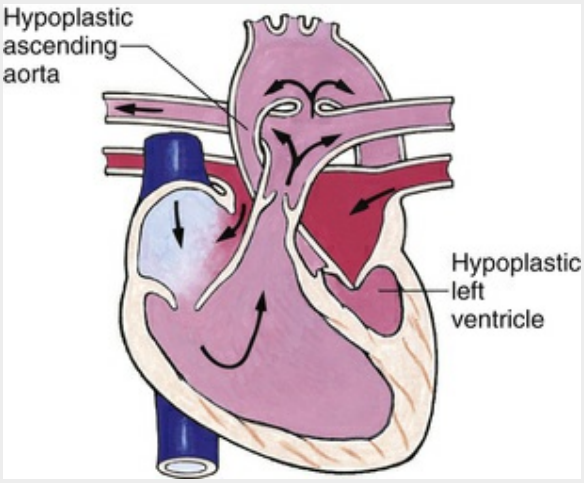

Description: Underdevelopment of the left side of the heart, resulting in a hypoplastic left ventricle and aortic atresia. Most blood from the left atrium flows across the patent foramen ovale to the right atrium, to the right ventricle, and out the pulmonary artery. The descending aorta receives blood from the PDA supplying systemic blood flow.

Pathophysiology: An ASD or patent foramen ovale allows saturated blood from the left atrium to mix with desaturated blood from the right atrium and to flow through the right ventricle and out into the pulmonary artery. From the pulmonary artery, the blood flows both to the lungs and through the ductus arteriosus into the aorta and out to the body. The amount of blood flow to the pulmonary and systemic circulations depends on the relationship between the pulmonary and systemic vascular resistances. The coronary and cerebral vessels receive blood by retrograde flow through the hypoplastic ascending aorta.

Clinical manifestations: The patient has mild cyanosis and signs of HF until the PDA closes and then progressive deterioration with cyanosis and decreased cardiac output, leading to cardiovascular collapse. The condition is usually fatal in the first months of life without intervention.

Therapeutic management: Neonates require stabilization with mechanical ventilation and inotropic support preoperatively. A