enters the common trunk so that pulmonary and systemic circulations are mixed. Blood flow is distributed to the pulmonary and systemic circulations according to the relative resistances of each system. The amount of pulmonary blood flow depends on the size of the pulmonary arteries and the pulmonary vascular resistance. Generally, resistance to pulmonary blood flow is less than systemic vascular resistance, which results in preferential blood flow to the lungs. Pulmonary vascular disease develops at an early age in patients with truncus arteriosus.

Clinical manifestations: Most infants are symptomatic with moderate to severe HF and variable cyanosis, poor growth, and activity intolerance. There is a holosystolic murmur at the left sternal murmur with a diastolic murmur present if truncal regurgitation is present. Thirty-five percent of patients have 22q11 deletions (Goldmuntz, Clark, Mitchell, et al, 1998).

Surgical treatment: Early repair is performed in the first month of life. It involves closing the VSD so that the truncus arteriosus receives the outflow from the left ventricle and excising the pulmonary arteries from the aorta and attaching them to the right ventricle by means of a homograft. Currently, homografts (segments of cadaver aorta and pulmonary artery that are treated with antibiotics and cryopreserved) are preferred over synthetic conduits to establish continuity between the right ventricle and pulmonary artery. Homografts are more flexible and easier to use during the procedure and appear less prone to obstruction. Postoperative complications include persistent heart failure, bleeding, PAH, dysrhythmias, and residual VSD. Because conduits are not living tissue, they will not grow along with the child and may also become narrowed with calcifications. One or more conduit replacements will be needed in childhood.

Prognosis: Mortality is greater than 10%; future operations are required to replace the conduits.

Hypoplastic Left Heart Syndrome