

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome occurs when the left heart structures are underdeveloped, resulting in a left ventricle that cannot adequately support systemic cardiovascular output. As a result, the patient is left with essentially one functional ventricle (the right ventricle which normally pumps blood out to the lungs) that must be "rerouted and reconfigured" to pump blood to the body. Blood is directed to the lungs by passive flow as opposed to pump flow. This is done through a series of operations throughout the first 2 years of a child's life:

- *Norwood* (usually done in the first days of life) - combines the pulmonary artery and aorta to direct flow coming back from the body and lungs back out to the body. An extra connection to the lungs is also made at this time to allow for adequate oxygenation
- *Bidirectional Glenn* (done at 4 to 6 months) - The superior vena cava, which normally returns deoxygenated blood from the head and upper extremities back to the heart, is connected directly to the right pulmonary artery.
- *Fontan* (done at 18 months to 2 years) - The inferior vena cava, which returns blood from the lower extremities, liver, and kidneys, is connected via a conduit or baffle to the superior vena cava, which is already connected to the pulmonary arteries. At this point, ALL systemic venous return is going to the lungs, is oxygenated, returned to the pulmonary venous atrium to the single ventricle and out toward the body.