

Hypoplastic left heart syndrome is a term used to describe a group of closely related rare heart defects that are present at birth (congenital). The normal heart has four chambers. The two upper chambers, known as atria, are separated from each other by a fibrous partition known as the atrial septum. The two lower chambers are known as ventricles and are separated from each other by the ventricular septum. Valves connect the atria (left and right) to their respective ventricles. The valves allow for blood to be pumped through the chambers. Blood travels from the right ventricle through the pulmonary artery to the lungs where it receives oxygen. The blood returns to the heart through pulmonary veins and enters the left ventricle. The left ventricle sends the now oxygen-filled blood into the main artery of the body (aorta). The aorta sends the blood throughout the body. Hypoplastic left heart syndrome is a rare disorder that affects males (67 percent) more often than females. The estimated prevalence of the disorder is 1 in 100,000 live births. Hypoplastic left heart syndrome accounts for 7-9 percent of all congenital heart defects. The symptoms of this disorder are present at birth (congenital). The diagnosis of hypoplastic left heart syndrome is made based upon a thorough clinical evaluation, identification of characteristic findings and a variety of specialized tests. Such tests used to confirm hypoplastic left heart syndrome in newborns include x-ray examination and a special ultrasound test to study the structure and function of the heart (echocardiography).