What Is It?

Hypoplastic Left Heart Syndrome (HLHS) is characterized by multiple small (hypo means small) structures on the left side of the heart.

The hypoplastic structures include the mitral valve (1 in diagram below), aortic valve (2), and the left ventricle itself (3). In addition, the ascending aorta (4) is abnormally narrow.
What Are Its Effects?

In Hypoplastic Left Heart, blood flow to the body is severely restricted. After birth, a baby with Hypoplastic Left Heart Syndrome will be ill and have poor color. In some cases where the Ductus Arteriosus remains open (or patent) - known as the Patent Ductus Arteriosus (PDA), there may be surprisingly few initial symptoms.

When the Patent Ductus Arteriosus (PDA) closes, the infant enters profound shock. Without treatment, the infant will generally die within a few weeks of birth.

How Is It Treated?

Medications may help a patient with Hypoplastic Left Heart to some degree, but surgical management (a Norwood Procedure or a heart transplant), will be necessary within a few weeks of birth.

In the Norwood Procedure (see the diagram below), a major blood vessel to the body (aorta) is constructed from the base of the pulmonary artery and the narrowed aorta.

In addition, a small tube made of Gore-Tex (known as a Modified Blalock-Taussig Shunt), shown in yellow on the diagram, is inserted between the branches of the pulmonary artery (PA) and a branch of the aorta to ensure blood flow to the lungs after the PDA closes.
Hypoplastic Left Heart Syndrome – Circulation after a Norwood Procedure