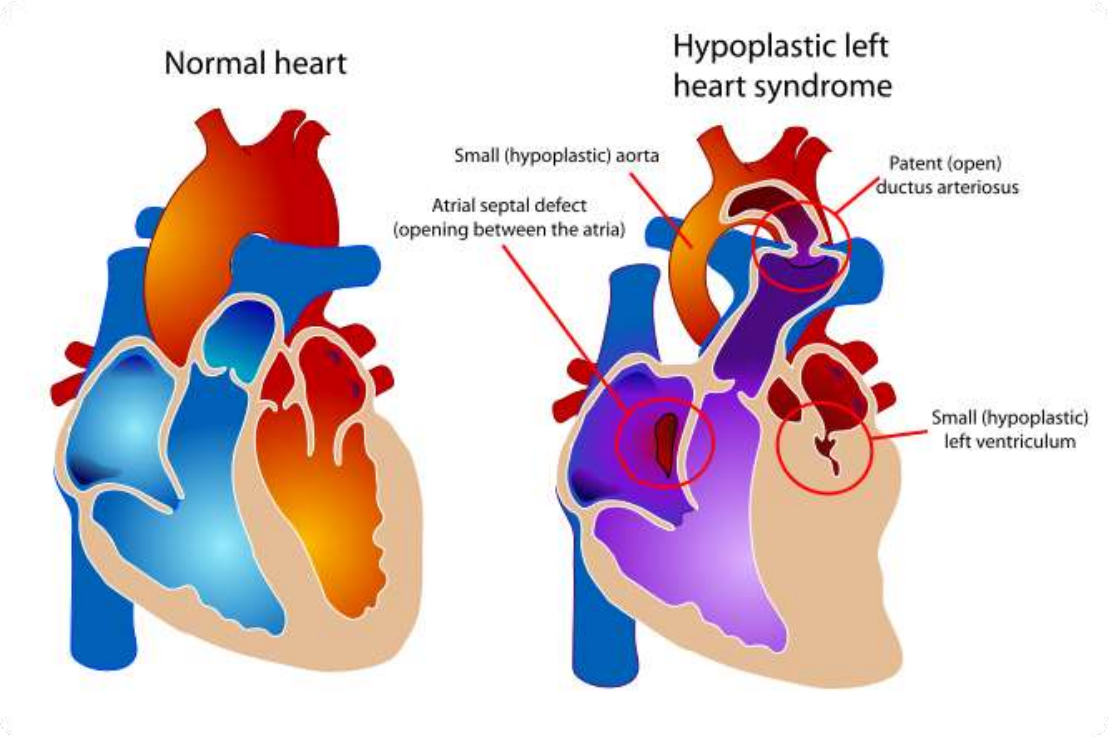


Hypoplastic Left Heart Syndrome

Definition:

In Hypoplastic left heart syndrome (HLHS), the entire left side of the heart (left atrium, left ventricle, aorta, mitral valve and aortic valve) is not well developed.

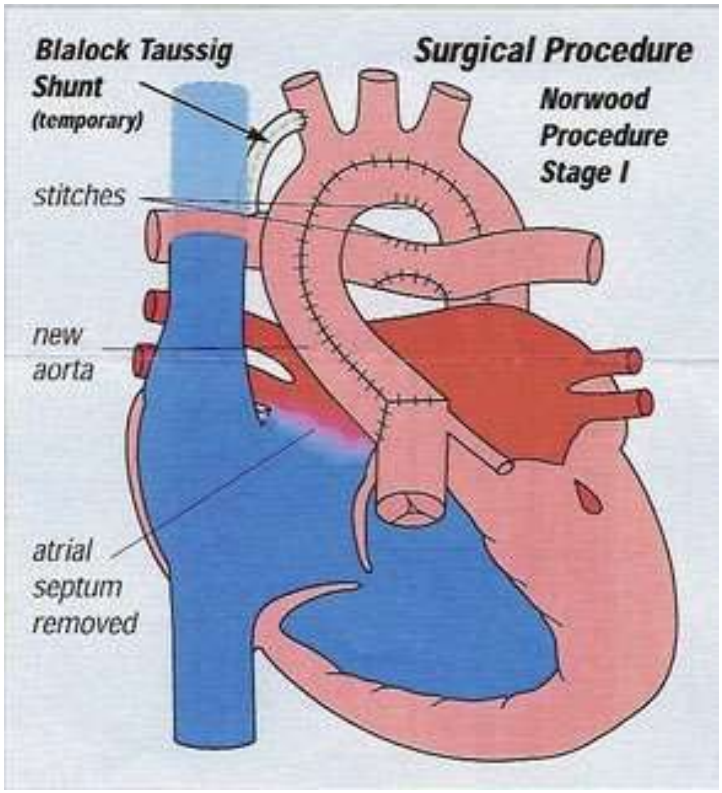
What causes it?

Cause is unknown, however it is known to be associated with other heart defects.

How does it affect the heart?

In HLHS, blood returning from the lungs to the left side of the heart (via pulmonary veins) cannot flow through the left side of the heart. Instead, they will go through a hole between the upper chambers (atria) to the right side where the blood is pumped into the pulmonary artery. The blood eventually reaches the aorta again via a patent ductus arteriosus (a connection between the pulmonary artery and aorta). At birth, the baby will seem normal but immediately deteriorates in condition within a few days of birth, as the patent ductus arteriosus (PDA) closes. The baby will have rapid feeding difficulty and rapid breathing and will look severely ill as the body is not receiving oxygen. If not treated, the condition will lead to death within a few days up to a month.

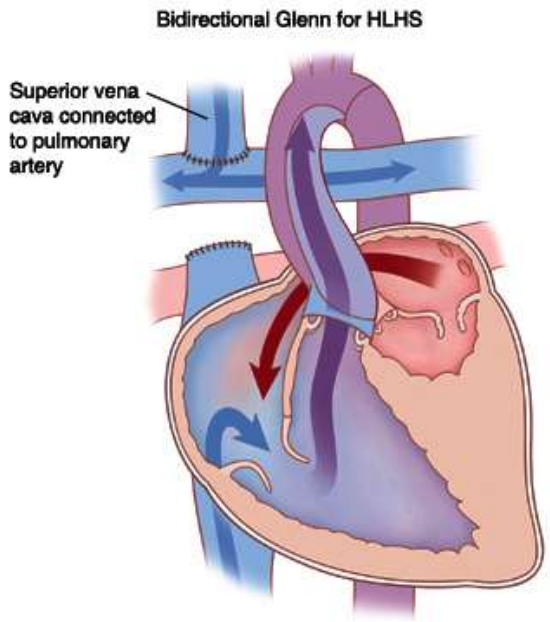
Management options:

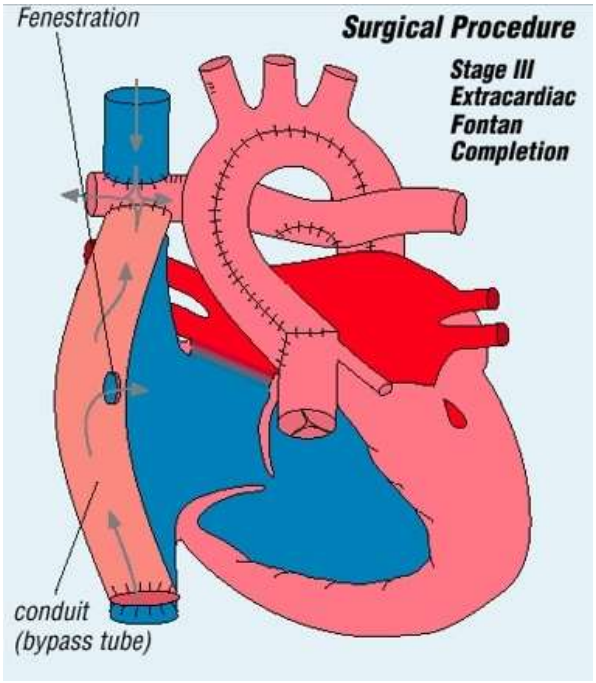


The problem cannot be corrected 100%, but with a series of operations called Norwood operation or heart transplantation, it can be treated. Until the child is ready for surgery, the PDA must remain open through use of medication. The surgery is very complex and due to its complexity and the risks it carries, the surgery must be discussed at length with the family and the doctor.

The first surgery to be done is known as a first stage Norwood procedure. This allows the right ventricle to pump blood to the aorta (which supplies the whole body except the lung) from where blood should flow to the lung arteries via a shunt created by the surgeon known as a Sano shunt or a Blalock- Taussig shunt (BT shunt). The Norwood operation must be carried out as soon as possible after birth.

If successful, the next step will be a bidirectional Glenn (2^{nd} stage Norwood) that is performed between the ages of 4 months and 12 months and the third stage known as a Fontan will be done between 18 months and 3 years. The second and third operations basically create a single ventricle in the patient that is capable of pumping only oxygenated blood to the body via the aorta and deoxygenated blood to the lungs via the pulmonary artery which receives the blood directly from the systemic veins without passing through the heart pump as the normal circulation.





Some doctors will discuss the potential for a heart transplant, as it provides the child with a normal heart, although they will need to be on medication to prevent the body's rejection to the new heart for life, and other transplant related complications may occur.

What activities can my child do?

There will definitely be limiting of physical activity and involvement in strenuous sports as dictated by your pediatric cardiologist.

Follow up in future?

There will be a lifelong follow up with your pediatric cardiologist as the doctor will need to check how the heart is working and monitor doses of medication and the need for further surgeries.

Children may need to receive antibiotics before any surgical or dental procedures to prevent infective endocarditis.