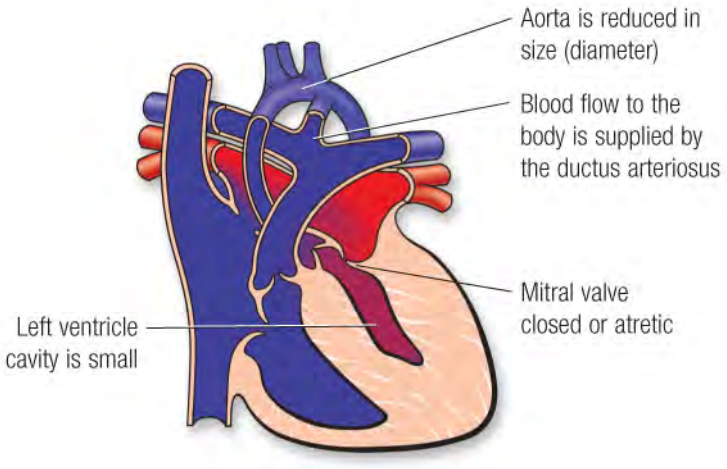
Hypoplastic Left Heart Syndrome

What is it?

In hypoplastic left heart syndrome (HLHS), the heart's left side — including the aorta, aortic valve, left ventricle and mitral valve — is underdeveloped.



What causes it?

In most children, the cause isn't known. Some children can have other heart defects along with HLHS.

How does it affect the heart?

In HLHS, blood returning from the lungs must flow through an opening in the wall between the atria (atrial septal defect). The right ventricle pumps the blood into the pulmonary artery and blood reaches the aorta through a patent ductus arteriosus (see diagram).

How does the defect affect my child?

The baby often seems normal at birth but comes to medical attention within a few days of birth as the ductus closes. The baby may appear ashen, have rapid and difficult breathing and have difficulty feeding. This heart defect is usually fatal within the first days or month of life unless it's treated.

What can be done about the defect?

This defect isn't correctable, but some babies can be treated with a series of operations, or heart transplantation. Until an operation is performed, the ductus is kept open by intravenous medication. Because these operations are complex and need to be adapted for each child, it's necessary to discuss all the medical and surgical options with your child's doctor.

If you and your child's doctor agree that surgery should be performed, it will be done in several stages. The first stage, referred to as the Norwood procedure, allows the right ventricle to pump blood to both the lungs and the body without the need for the ductus to be kept open. Blood is directed to the lungs through either a Blalock-Taussig (arrow on inserted picture) or Sano shunt. The Norwood procedure must be performed soon after birth.

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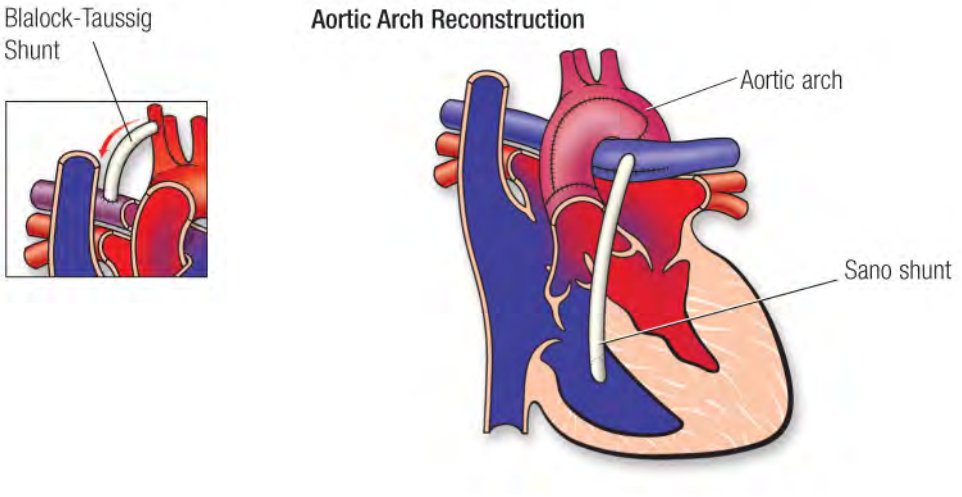
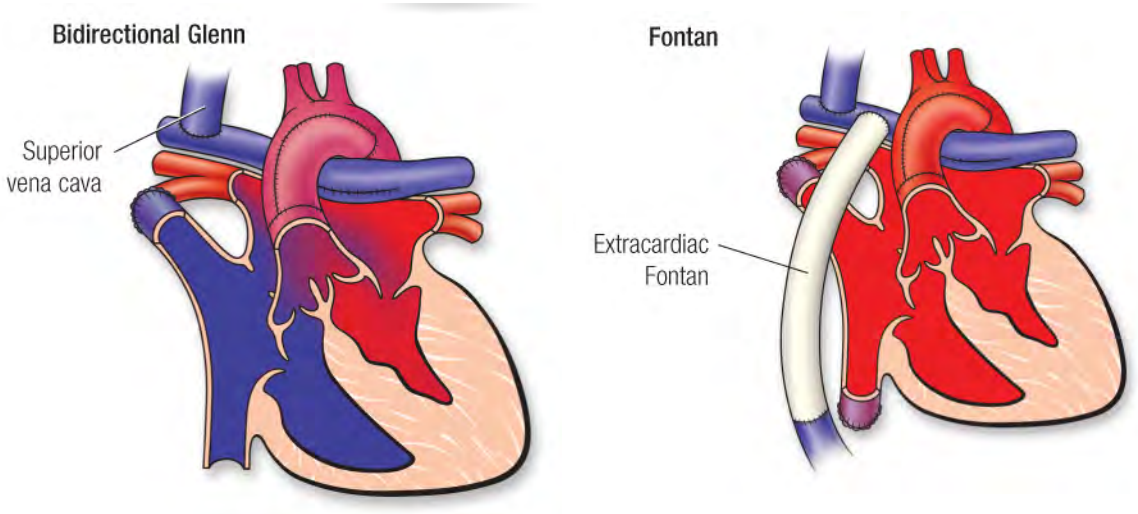


The second stage (bidirectional Glenn or hemi-Fontan) is usually performed between 4 and 12 months and the third stage (lateral tunnel Fontan or extracardiac Fontan) is usually performed between 18 months and 3 years.



These operations create a connection between the veins returning low-oxygen (bluish) blood to the heart and the pulmonary artery. The goal is to allow the right ventricle to pump only oxygenated blood to the body and to prevent or reduce cyanosis (lower than normal blood oxygen levels). Some infants require several intermediate operations to achieve this.

Some doctors recommend heart transplantation to treat HLHS. Although it can provide the infant with a heart that has normal structure, the infant will require life-long medications to prevent rejection. Many other transplant-related problems can develop, and these should be discussed with your child's doctor.

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What activities can my child do?

Children with HLHS may be advised to limit their physical activities to their own endurance. Generally, many competitive sports pose greater risk. Your child's pediatric cardiologist will help determine the proper level of activity.

What will my child need in the future?

Children with HLHS require lifelong follow-up by a cardiologist for repeated checks of how their heart is working. Virtually all children with HLHS will require heart medicines, heart catheterization and additional surgery.

What about preventing endocarditis?

Children with HLHS are at increased risk for developing endocarditis. Ask your pediatric cardiologist about your child's need to take antibiotics before certain dental procedures to help prevent endocarditis.