

Hypoplastic Left Heart Syndrome

BIRTH DEFECT RESEARCH FOR CHILDREN



What is Hypoplastic Left Heart Syndrome?

A baby with Hypoplastic Left Heart Syndrome has an underdeveloped left ventricle, the heart chamber that pumps blood through the aorta to the entire body. In some cases the mitral valve (which connects the atrium and ventricle), the aortic valve (which connects the heart to the major vessels leading from the lungs), and the ascending aorta (which carries blood from the heart to be distributed through the body) may be absent or underdeveloped. These defects prevent the left side of the baby's heart from functioning properly. Consequently, the right side of the baby's heart takes on the function of pumping blood to the body through the ductus arteriosus into the aorta. The ductus arteriosus connects the pulmonary artery to the aorta and allows the blood to bypass the lungs during fetal development since the baby does not breathe until after birth. It normally closes shortly after birth. When the ductus arteriosus closes in a baby with Hypoplastic Left Heart Syndrome, the blood supply to the baby's body is cut off.



Hypoplastic Left Heart Syndrome



How many children have Hypoplastic Left Heart Syndrome?

Hypoplastic Left Heart Syndrome occurs about 1 in 10,000 births. It is the fourth most common heart defect in newborns and is the most common cause for early cardiac death (usually within the first week of life).

How do you know if your child has Hypoplastic Left Heart Syndrome?

Hypoplastic Left Heart Syndrome can be diagnosed through obstetrical ultrasounds and on fetal echocardiograms. If undiagnosed during pregnancy, the baby's condition will become noticeable within the first hours or days of life. The baby may seem normal but can soon develop respiratory distress and his or her extremities may appear pale and cool. As the ductus arteriosus closes, the right ventricle will no longer be able to pump blood to the aorta, the baby's skin will turn blue, and the baby will experience shock and multi-organ failure. If untreated, the heart defect will become fatal within the first days or weeks of the baby's life.

What causes Hypoplastic Left Heart Syndrome?

The cause of Hypoplastic Left Heart Syndrome is unknown. Go to <http://birthdefects.blogspot.com/> for the latest research on the causes of congenital heart defects.

How can you help a child with Hypoplastic Left Heart Syndrome?

If Hypoplastic Left Heart Syndrome is diagnosed during pregnancy, plans can be made for the stabilization of the newborn with respiratory treatments and medications at the time of birth. Intravenous medication will be used to keep the ductus arteriosus open. Treatment options include heart transplantation and reconstructive surgery. The baby's condition may dictate which procedure is performed. Heart transplantation is limited due to the low availability of donor hearts. Therefore, the most common treatment is staged reconstruction that typically involves three

surgeries. The first operation, which is typically performed in the first week of life, is called the Norwood Procedure. This operation reconstructs the aorta and pulmonary arteries, so the blood can flow from the right ventricle to the lungs and the aorta. The second surgery, called the Bi-directional Glenn Procedure, is typically performed at 4-6 months. The final surgery, the Fontan, is typically done at 1-3 years. These last two surgeries reconstruct the heart so blood flows only from the right ventricle to the body and then from the body to the lungs.

What's in the future for a child with Hypoplastic Left Heart Syndrome?

Heart transplants and the three-stage surgical procedures are extremely complex surgeries. Hospitals that have a history of performing many of these surgeries will generally have a higher success rate. The survival rate for infants undergoing the Norwood Procedure is 75% or better. This is the most complex and highest risk procedure of the three operations. The survival rate for children undergoing the Glenn and Fontan operations is 90% or better. All children with Hypoplastic Left Heart Syndrome will require lifelong follow-up for cardiac evaluation. Most will require some continuing cardiac medications. Transplant patients will need to take anti-rejection medications for the rest of their lives.

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