Choosing Your Child’s Care

When you consider Cleveland Clinic Children’s for heart care, from routine visits to the most complex procedures, you are in the very best hands. Our team of pediatric heart surgeons, pediatric cardiologists, cardiac anesthesiologists, intensive care unit (ICU) physicians and specially trained pediatric cardiac nurses is among the most well-trained and knowledgeable in the world. Plus, our pediatric heart program is continually ranked among the nation’s best by U.S. News & World Report.

USING THIS GUIDE

This guide provides an overview of what you need to know about hypoplastic left heart syndrome, and how it is diagnosed and treated.

Remember, it is every parent’s right to ask questions and seek a second opinion. Cleveland Clinic Children’s specialists are available for consultation if you have a question about your child.
Congenital heart disease: A better outlook

The heart is one of the body’s most powerful organs, circulating blood that carries oxygen and nutrients to every inch of the body.

The heart forms at eight to 10 weeks of development. In about eight infants out of 1,000, one or more of the heart’s structures fails to develop properly. This is called congenital heart disease. Whether symptoms appear at birth, during childhood or later on, it is always hard for parents to hear these words.

The good news is that congenital heart disease is diagnosed earlier and treated more successfully today than ever before. Now, babies with congenital heart disease commonly grow well into adulthood with the intervention of pediatric heart experts.

About hypoplastic left heart syndrome

In hypoplastic left heart syndrome, or HLHS, the left side of the heart is underdeveloped. This syndrome affects one to two infants out of 10,000 and represents about 1 percent of all congenital heart disease. HLHS is slightly more common in boys than in girls. In rare cases, a defective gene may be inherited, but in most cases, HLHS has no clear cause.

How the anatomy of the heart is affected

In HLHS, four heart structures on the left side of the heart are typically underdeveloped:

Mitral valve: the valve that allows blood from the left atrium (upper chamber) to flow into the left ventricle (lower chamber)

Left ventricle: the main pumping chamber of the heart

Aortic valve: the valve that allows the left ventricle to pump blood out into the aorta

Aorta: the main blood vessel from the heart, which distributes blood throughout the body to nourish organs and tissues
Normal circulation in the womb

Before birth, the fetus needs oxygen from the mother’s circulation to nourish vital organs and tissues. That is because the lungs don’t start working until the baby is born. Normally, a blood vessel called the ductus arteriosus remains open, or patent, in the fetal heart. This allows the mother’s blood to bypass the fetal lungs and the left side of the heart. This “patent ductus arteriosus,” or PDA, allows growth and development to continue.

Once the baby is born and begins to breathe air, he or she no longer gets oxygen through the mother’s circulation. Within a few hours, the PDA begins to close, leaving the left side of the baby’s heart responsible for delivering oxygen-rich blood to the body.

Circulation in HLHS

In HLHS, this natural sequence of events does not take place. If the PDA were allowed to close normally, the underdeveloped left ventricle and aorta would not be able to take over. Blood flow would be insufficient to meet the baby’s needs without medical and surgical intervention. That is why medications are used to keep the PDA from closing while the newborn awaits surgery.

How HLHS is diagnosed

HLHS is usually diagnosed in the womb. Serious heart abnormalities are usually detected by an obstetrician during a fetal ultrasound. The obstetrician then refers the mother to a pediatric cardiologist.

The pediatric cardiologist will perform a more specialized ultrasound test called a fetal echocardiogram (a safe, simple, noninvasive technique that uses sound waves to create a moving picture of the heart’s internal structures and blood flow). This is when a diagnosis of hypoplastic left heart syndrome is made.

After birth, the diagnosis is confirmed with another echocardiogram and possibly other tests so that treatment can be started immediately. Newborns not treated promptly for HLHS can turn pale or develop blue-colored skin, lips and nail beds; have sweaty, clammy or cool skin; experience breathing difficulties and feeding problems; and develop a rapid heart beat.

Managing HLHS before birth

Because of the PDA, good prenatal care and close follow-up with a pediatric cardiologist are usually all the mother needs before the baby is born. This is because the fetus is not yet using the underdeveloped left side of the heart.

However, in rare cases, the hole that normally exists in the atrial septum (wall between the upper chambers of the heart) is too small or closed off. Cleveland Clinic Children’s heart specialists may use a special fetal catheter technique to open the atrial septum in order to avoid severe lung damage and improve the chances of survival.
Treating HLHS after your baby is born

**Medicine at birth**

Newborns with HLHS are given an intravenous (IV) medication to keep the ductus arteriosis open. This allows the PDA to continue to bypass the small left heart, relying instead on the larger and stronger right heart. The medication allows blood to be delivered throughout the body until surgery can be performed.

**Surgery during infancy and childhood**

Children with HLHS need a series of operations to reroute blood flow. The standard operations used are named for the surgeons who developed them: the Norwood procedure, the Glenn procedure and the Fontan procedure. They are also simply referred to as Stage 1, Stage 2 and Stage 3 surgery for HLHS.

**THE NORWOOD OPERATION (STAGE 1)**

This is the most complex of the three operations. It is typically performed in the first week of life. The goal of the Norwood procedure is to let the right ventricle become the sole pumping chamber of the heart, delivering blood to both the body and the lungs. Three major steps are involved:

- The small aortic valve and ascending aorta are combined with the normal pulmonary valve. A patch is used to reconstruct a new, larger aorta.

- The wall between the two atria, or atrial septum, is removed so that all the blood from the right and left atria can reach a single pumping chamber: the right ventricle.

- A small tube, or shunt, made of synthetic material is connected from the right ventricle to the pulmonary arteries. This lets the right ventricle pump blood to the lungs so that it receives oxygen.

The Norwood operation is a major procedure with significant risks. Parents should discuss it in detail with their pediatric cardiologist and pediatric cardiac surgeon.

**THE GLENN OPERATION (STAGE 2)**

Stage 2 surgery is performed when the lungs are more mature, typically three to six months after Stage 1 surgery. The risks of mortality and complications are significantly lower for the Glenn operation than they are for the Norwood operation.

At this time, the baby’s lungs have begun to relax, allowing blood to flow more easily from the heart to the lungs and setting the stage for the Glenn operation. The Glenn operation is designed to ease the workload of the single right ventricle, which will no longer need to do the extra work of pumping blood to the lungs. The Glenn operation involves:

- Removing the shunt placed during the Norwood operation.
• Disconnecting the large vein returning blood from the upper half of the body (superior vena cava) to the heart and connecting it to the pulmonary arteries. Blood from the upper half of the body will now go straight to the lungs, where it receives oxygen.

After this procedure, blood returning from the lower half of the body through the veins still goes directly to the heart without going through the lungs. That means the baby receives a mixture of oxygen-rich and oxygen-poor blood each time the heart sends out blood to the body. Although babies tolerate these below-normal levels of oxygenated blood well, their skin color may be a bit pale or slightly blue.

THE FONTAN OPERATION (STAGE 3)
Cleveland Clinic Children’s heart surgeons usually perform this procedure when children with HLHS are between the ages of 2 and 4. By this time, their lungs have matured enough to handle blood returning from both the upper and lower halves of the body.

The Fontan procedure involves connecting the major blood vessel that returns blood from the lower part of the body (inferior vena cava) to the pulmonary artery using a large synthetic tube.

After this operation, blood will flow to the lungs from both the upper and lower body. The blood that the heart is able to pump throughout the body is now fully oxygenated, and the child’s skin color becomes more pink.

AN ALTERNATIVE: THE HYBRID PATHWAY
The hybrid pathway is an alternative treatment for HLHS that combines catheter intervention with surgery to reroute blood flow. It still involves three operations, but the second stage is the most complex.

The first stage involves inserting a stent through a catheter into the PDA to keep it from closing and surgically narrowing the pulmonary arteries to prevent blood flow from overwhelming the lungs.

The second stage combines the Norwood operation (aortic reconstruction) and Glenn operation (connection of the superior vena cava to the pulmonary arteries). It is performed when infants are 3 to 6 months old.

The third stage involves performing the Fontan operation, described above.

The risks associated with the hybrid pathway are likely to be similar to the risks associated with the standard Norwood pathway. Research continues to evaluate the long-term risks and benefits of the hybrid pathway.

HEART TRANSPLANTATION
At most major U.S. centers, the results of the Norwood procedure are superior to the results of heart transplantation for infants with HLHS. One reason is that these babies cannot survive the long wait for a donor heart. Heart transplantation is generally reserved for infants with complications of HLHS who are unlikely to survive the Norwood procedure.
Deciding what’s best for your child

HLHS and its treatments are extremely complex because each child with HLHS is different. As a result, the outlook for children with HLHS varies considerably.

Ultimately, choosing the best surgical approach will depend upon your baby’s overall health and unique heart anatomy, and on your wishes as parents. Talk to your heart specialist about your baby’s outlook and the best medical and/or surgical approach for your child. You also have the right to ask about the frequency and results of HLHS operations performed at any hospital.

Following children as they grow

All children with HLHS require lifelong follow-up with a pediatric cardiologist to monitor their progress. Most children need to take heart medicines and some may need further surgery later in life. However, the majority of children with HLHS are able to enjoy active, happy lives.

Fetal and newborn heart care

Our physicians not only diagnose heart disease in children, we can determine management strategies before they are even born.

Complicated and high-risk pregnancies are expertly managed within our Level III perinatal center.

Critically ill newborns receive the highest level of care available in our Level III neonatal intensive care unit.

Leaders in research and innovation

Our surgeons developed and perfected the double switch operation for complex transposition of the great arteries. We also pioneered devices to close holes between the chambers of the heart.

Our physician investigators are sponsored by corporate and government organizations with the goal of advancing treatment for pediatric patients with congenital heart defects.

Lifetime care for children and adults

We are one of the few heart centers in Ohio dedicated to the lifelong care of children and adult patients with congenital heart defects, including post-surgical care and treatment.

We work in close collaboration with the world-renowned Cleveland Clinic Heart & Vascular Institute to ensure that children and adults receive the best possible care with leading-edge technology and clinical expertise.

Pediatric nutrition support team

Nutrition is a critical component to therapy for young children with chronic diseases and medical conditions. Proper nutritional support prevents malnutrition, guides appropriate physical growth and development, and helps to maintain a strong immune system.
A pediatric registered dietitian is a licensed health professional who specializes in the nutritional care of children and understands their specific nutritional needs. Cleveland Clinic Children’s has a team of registered dietitians, board-certified in pediatrics, who collaborate with physician teams to provide expert, evidence-based nutritional care for infants, children and adolescents.

The Pediatric Nutrition Support Team provides individual counseling to assess each child’s nutritional needs, develop the most appropriate plan of nutritional support, and monitor nutritional status to ensure an optimal outcome.

**Additional information**
Meet our cardiologists and heart surgeons and learn more at clevelandclinicchildrens.org/hlhs

**Patient Services**

**MyChart.Caregiver**
Parents and caregivers of children under age 18 can view test results, renew prescriptions, review past appointments and request new ones using Cleveland Clinic MyChart.Caregiver®. This secure online healthcare management tool connects parents and legal guardians to portions of a child’s medical record at any time, day or night. Parents visit clevelandclinic.org/mychart to register.

**Medical Concierge**
For complimentary assistance for out-of-state patients and families, call 800.223.2273, ext. 55580, or email medicalconcierge@ccf.org.

**Global Patient Services**
For complimentary assistance for international patients and families, please call the International Patient Access Office at +1.216.444.8184. If you are calling from inside the U.S., call toll free at 866.320.4573. You can also visit clevelandclinic.org/gps.

**Need a second opinion, but cannot travel to Cleveland?**
Our MyConsult® service offers secure online second opinions for patients who cannot travel to Cleveland. Through this service, patients enter detailed health information and mail pertinent test results to us. Cleveland Clinic experts then render an opinion that includes treatment options or alternatives and recommendations regarding future therapeutic considerations.

To learn more about MyConsult, visit clevelandclinic.org/myconsult.

**Excellent outcomes at Cleveland Clinic Children’s**
Our Center for Pediatric and Congenital Heart Diseases manages infants, children and young adults with all types of inherited and acquired heart problems. We routinely perform complex operations and heart catheterizations not done elsewhere, and we use the least invasive techniques whenever possible.

Infants and children are referred from across the country and around the world to Cleveland Clinic Children’s. Our outcomes for HLHS surgery are significantly better than the national average. See this year’s Cleveland Clinic Children’s Outcomes book for data on our treatment of pediatric and congenital heart disease at Cleveland Clinic.

Call 216.445.5015 for a same-day appointment or visit clevelandclinicchildrens.org
Cleveland Clinic Children's Locations
Cleveland Clinic Children's 300+ pediatricians and pediatric subspecialists offer comprehensive medical, surgical and rehabilitative care at more than 40 community locations throughout Northeast Ohio. Find one near you.

Cleveland Clinic Children's Staff
Please visit clevelandclinicchildrens.org/staff for a complete listing.

Cleveland Clinic Children's
9500 Euclid Ave., Cleveland, OH 44195

Cleveland Clinic Children's provides comprehensive medical, surgical and rehabilitative care for infants, children and adolescents. Our more than 300 pediatric physicians accommodate more than 750,000 outpatient visits and 18,000 inpatient admissions per year at the children's hospital and outpatient facilities on our main campus, at the Cleveland Clinic Children's Hospital for Rehabilitation campus, and at regional hospitals, family health centers and other locations across Northeast Ohio. Cleveland Clinic Children's & Pediatric Institute is one of 27 institutes at Cleveland Clinic designed to offer highly integrated care and conduct innovative research across multiple settings. It is backed by the full resources of Cleveland Clinic, a nonprofit, academic medical center ranked among the nation's top hospitals (U.S. News & World Report).
clevelandclinicchildrens.org

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