Jamie Swartzel
20 Claiborne Court
Florence, KY 41042
August 22, 2013

Dr. Dorothy Perkins
Superintendent
Gallatin County Schools
75 Boardwalk
Warsaw, KY 41095

Dear Dr. Dorothy Perkins:

I am requesting maternity leave from the Gallatin County School District from November 25, 2013 until April 6, 2014 with the impending arrival of Jeffrey Thomas Swartzel. My due date is Thursday, December 5, 2013 but I am under the watchful eye of my obstetrician with having a high risk pregnancy.

I am requesting an extension of my maternity leave because Jeffrey Thomas has already been diagnosed with Hypoplastic Left Heart Syndrome. He will have a series of two surgeries prior to being a year old. The first surgery will be completed within the first week to ten days of his life, while the second surgery will be performed when he is three to seven months of age.

Sincerely,

Jamie Swartzel

Jamie Swartzel
7th and 8th Grade Mathematics Teacher
Gallatin County Middle School

Heart Institute Encyclopedia

Cincinnati Children's

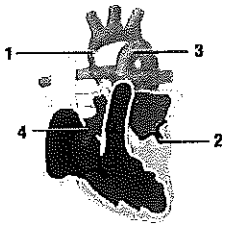
Hypoplastic Left Heart Syndrome / Norwood Operation

Hypoplastic Left Heart Syndrome (HLHS) is one of the most complex cardiac defects seen in the newborn and remains probably the most challenging to manage of all congenital heart defects. It is one of a group of cardiac anomalies that can be grouped together under the description "single ventricle" defects.

Glossary

Flash movie Graphic summary

Example of a normal heart



Example of Hypoplastic Left Heart Syndrome

1. Hypoplastic ascending aorta and aortic arch.
2. Hypoplastic left ventricle.
3. Large patent ductus arteriosus supplying the only source of blood flow to the body.
4. Atrial septal defect allowing blood returning from lungs to reach the single ventricle.

In a child with Hypoplastic Left Heart Syndrome, all of the structures on the left side of the heart (the side which receives oxygen-rich blood from the lungs and pumps it out to the body) are severely underdeveloped.

The mitral and aortic valves are either completely "atretic" (closed), or they are very small. The left ventricle itself is tiny, and the first part of the aorta is very small, often only a few millimeters in diameter.

This results in a situation where the left side of the heart is completely unable to support the circulation needed by the body's organs, though the right side of the heart (the side that delivers blood to the lungs) is typically normally developed.

Blood returning from the lungs to the left atrium must pass through an atrial septal defect (ASD) to the right side of the heart.

The right ventricle must then do a "double duty" of pumping blood both to the lungs (via the pulmonary artery) and out to the body (via a patent ductus arteriosus (PDA)). The patent ductus arteriosus, a normal structure in the fetus, is often the only pathway through which blood can reach the body from the heart. When the ductus arteriosus begins to close, as it typically does in the first days of life, the blood flow to the body will severely diminish resulting in dangerously low blood flow to vital organs and leading to shock. Without treatment, Hypoplastic Left Heart Syndrome is uniformly fatal, often within the first hours or days of life.

Signs and Symptoms

As mentioned above, infants with Hypoplastic Left Heart Syndrome can develop life-threatening shock when the ductus arteriosus begins to close. In most cases, however, the ductus arteriosus is widely open at the time of birth, supplying the blood flow to the body and babies may not be diagnosed right away. As the ductus arteriosus closes, which it typically will in most infants in the first hours or days of life, the perfusion to the body is seriously diminished and shock rapidly ensues.

Newborns with Hypoplastic Left Heart Syndrome will typically have lower-than-normal oxygen saturations. This is because all of the blood from the lungs (the oxygenated "red" blood) mixes together in the single right ventricle before being pumped out of the lungs and body. Cyanosis, therefore, may be the first clue to the presence of a serious underlying cardiac condition. Respiratory distress (difficult or fast breathing) is often present because the lungs will tend to receive an excessively large amount of blood flow. There is often no or just a faint murmur present in newborns with Hypoplastic Left Heart Syndrome.

The pulses may be very weak in all extremities on examination depending on flow through the ductus arteriosus. Lethargy, poor feeding, and worsening respiratory distress may be seen as the ductus arteriosus closes. Ultimately, severe shock resulting in seizures, renal failure, liver failure, and worsening cardiac function may develop. Whether these problems are reversible depends on both the severity and the duration of shock.

Diagnosis of Hypoplastic Left Heart Syndrome

This heart defect is one of the most readily diagnosed on fetal echocardiograms and is one of the most common cardiac defects picked up on screening obstetrical ultrasounds. Such early diagnosis of the anomaly allows for prompt intervention for stabilization at the time of birth so that severe shock may be avoided.

Planning to deliver such an infant at a hospital capable of aggressive newborn resuscitation is important in improving the chances for a good outcome.

Echocardiography is the principal method of diagnosing Hypoplastic Left Heart Syndrome. It can give detailed information of the anatomy of the various cardiac structures affected in Hypoplastic Left Heart Syndrome, as well as important information about the function of the right ventricle, the heart valves, the size of the atrial septal defect (important for blood mixing) and the size of the patent ductus arteriosus.

Cardiac catheterization is rarely used as part of the initial evaluation, with this heart defect due to the high risks in an often unstable newborn. Catheterization, though, does play an important role in the evaluation of the cardiopulmonary function and anatomy in older children with Hypoplastic Left Heart Syndrome while planning for later stages in the treatment.

Treatment of Hypoplastic Left Heart Syndrome

The management of the newborn with Hypoplastic Left Heart Syndrome can be divided into the initial stabilization period and the operative / post-operative period.

If the fetus has been diagnosed before delivery, stabilization measures are started immediately so the newborn does not become unstable. In newborns that are delivered and then suspected of having Hypoplastic Left Heart Syndrome, stabilization begins even while diagnostic tests are going on. The rapid stabilization of these infants must begin as soon as the diagnosis is suspected.

Catheters are placed, usually in the umbilical blood vessels, which allow medications to be given and blood to be obtained for testing. An infusion of prostaglandin, a medication that prevents the patent ductus arteriosus from closing, is begun, thus maintaining the pathway for blood to reach the body from the right ventricle.

Even though the infant may have low oxygen saturations, supplemental oxygen is avoided since it tends to promote more blood flow to the lungs which may steal blood flow from the body and place excessive demands on the already stressed single right ventricle.

Manipulations of medications and respiratory treatments (including possible mechanical ventilation) are performed to optimally balance the flow of blood to the body and the flow of blood to the lungs.

Close monitoring is essential to detect any organ dysfunction and maintain cardiopulmonary stability because infants with this anomaly may be very unpredictable and undergo quite sudden changes.

There are essentially three treatment options that have been proposed for children with Hypoplastic Left Heart Syndrome.

In the past, due to poor outcomes with available treatments at that time, no treatment was often recommended. Today it is rare that a family may choose not to treat a child with Hypoplastic Left Heart Syndrome, though in cases when the infant is unable to be satisfactorily stabilized no treatment may be advised.

Cardiac transplantation in the newborn period is performed as primary treatment for Hypoplastic Left Heart Syndrome at some centers in this country. While transplantation has the advantage of replacing the very abnormal heart of a child with Hypoplastic Left Heart Syndrome with one of normal structure, this treatment is limited by the scarcity of newborn organs available for transplantation and the life-long need for anti-rejection therapy. Additionally, although outcomes for transplantation continue to improve, and the incidence of rejection is lowest in patients transplanted as newborns, the average life span of the transplanted heart is limited (currently less than 15 years).

The most commonly pursued treatment for Hypoplastic Left Heart Syndrome is "staged reconstruction" in which a series of operations, usually three, are performed to reconfigure the child's cardiovascular system to be as efficient as possible despite the lack of an adequate left ventricle. These surgeries do not correct the lesion, and are instead considered "palliative".

The first operation in the staged approach is known as the Norwood operation and is typically performed in the first week of life. With the Norwood operation, the right ventricle become the systemic or main ventricle pumping to the body. A "new" or "neo" aorta is made from part of the pulmonary artery and the original, tiny aorta, which is reconstructed / enlarged to provide blood flow to the body. Finally, to provide blood flow to the lungs, a small tube graft is placed either from an artery to the lung vessels (called a modified Blalock-Taussig shunt) or from the right ventricle to the lung vessels (called a Sano modification). Because of the extensive reconstruction of the aorta that must be done, this operation is one of the most challenging heart surgeries in pediatrics.

The subsequent operations in the staged reconstruction plan are the bi-directional Glenn procedure, typically done at 3 to 6 months of age, and the Fontan operation, typically done in children older than 2 or 3 years. These operations are described in more detail in the Heart Encyclopedia chapter on "Single Ventricle Cardiac Anomalies."

Norwood Surgery

Results with staged reconstruction for children with The Norwood operation is the most complex and highest risk procedure in the sequence of staged reconstruction for Hypoplastic Left Heart Syndrome. Current management at major pediatric heart centers has resulted in survival rates of 75 percent or better.

The recovery period in the hospital following the Norwood operation is often unpredictable and complicated, averaging about 3 to 4 weeks. A small percentage of patients who leave the hospital may continue to experience significant problems in the first months of life.

Occasionally, the right ventricle does not function well following the Norwood operation and in some case, cardiac transplantation may need to be considered.

If a child with Hypoplastic Left Heart Syndrome reaches the time for the second stage (about 4 to 6 months of age) without major complications, the survival through the Glenn and Fontan operations are much better, exceeding 90 percent with current methods.

Almost all children with Hypoplastic Left Heart Syndrome will continue to need some cardiac medications to maximize the efficient function of their heart, and all will require regular periodic follow-up visits with their cardiologist to evaluate their cardiac function and detect late complications such as arrhythmias.

Adult and Adolescent Management

All adult patients who started out life with HLHS have had a Fontan procedure. This procedure made them pink, but they have only one pumping chamber in their heart. As they age, many problems can occur, and Fontan patients need careful and thoughtful surveillance throughout their lives by experts in complex congenital heart disease. Many Fontan patients will have abnormal heart rhythms that require treatment. Some will have low oxygen levels in their blood that may limit their physical abilities, and that may require the blockage of abnormal vessels that were allowing blue blood to mix with red blood. Some Fontan patients may have weakening of their pumping chamber or leakage of one or more valves. Perhaps 10 percent of these patients may develop an unusual condition called "protein losing enteropathy." This requires very careful management. Fontan patients may also develop problems with liver or kidney function, so this needs to be monitored as well.

Most women with a congenital heart disease history may be able to successfully carry one or more pregnancies. Women with complex forms of congenital heart disease should definitely receive counseling and assessment before becoming pregnant, and should have their pregnancy managed by a special maternal fetal medicine team skilled at managing pregnancy in women with other health conditions.

Learn more about the Adolescent and Adult Congenital Heart Disease Program.

Contact Us

Contact Cincinnati Children's Heart Institute

Research Studies / Clinical Trials

- Marfan Syndrome (MFS) Medication Study
- Medication Study For Children Having Heart Surgery

Last Updated: 03/2010