Hypoplastic Left Heart Syndrome
Medical and Surgical Management
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Hypoplastic left heart syndrome is one of the most severe congenital heart defects. It occurs in 0.016 to 0.036 percent of live births and makes up between 1.4 to 3.8 percent of congenital heart disease. It is associated with hypoplasia of the ascending and transverse aorta, stenosis or atresia of the aortic valve, stenosis or atresia of the mitral valve, and hypoplasia of the left ventricle.

It varies as to when a patient presents clinically with HLHS, depending on ductal patency and the degree of atrial level restriction. A quarter of infants present during the first 24 hours after birth. Most infants present after 48 hours with feeding difficulties and respiratory distress that can rapidly progress to congestive heart failure and shock. If unrecognized or untreated, this condition is uniformly fatal, accounting for 25 percent of infant cardiac deaths during the first week of life and 15 percent during the first month of life.

Until the 1990s, the treatment of HLHS was associated with surgical mortality rates approaching or exceeding 90 percent. Children’s Hospital of Wisconsin has the best-reported outcomes in the treatment of HLHS, with the lowest-reported mortality rates (less than 6 percent) for the critical first-stage Norwood procedure. We have achieved these results through teamwork and innovations in prenatal diagnosis, neonatal management, surgical innovation, postoperative monitoring, and home monitoring following discharge.

Diagnosis and neonatal management
The majority of HLHS cases in Wisconsin are diagnosed prenatally, allowing for planned delivery at a center of excellence and the immediate initiation of prostaglandins to maintain patency of the ductus arteriosus. Diagnosis is often made when a prenatal ultrasound indicates a hypoplastic left ventricle. Following delivery, postnatal echocardiography is essential to confirm the diagnosis and to make observations on several critical anatomic points, including the following:

- **Atrial septum**: An intact or restrictive atrial septum results in obligatory obstruction to pulmonary venous return, wet lungs, and moderate to severe hypoxia requiring emergent intervention with atrial septectomy.
- **Aorta**: Severely hypoplastic aortas can be as tiny as 1 millimeter, essentially the size of a neonatal coronary artery, and result in reconstructive challenges at the time of the Norwood operation.
- **Tricuspid valve**: The tricuspid valve will serve as the systemic AV valve and can be associated with insufficiency, requiring repair at the time of the Norwood operation.

Preoperative management
HLHS results in a parallel circulation with each cardiac contraction ejecting blood from the right ventricle to both the lungs and the systemic circulation via the patent ductus arteriosus. Following initial diagnosis and stabilization with prostaglandin E1, newborns with HLHS are managed with therapies directed at balancing systemic and pulmonary circulation, critically avoiding pulmonary overcirculation and systemic underperfusion as the pulmonary vascular resistance drops.

Children’s Hospital of Wisconsin has championed the consistent use of two-site NIRS monitoring of tissue oxygen saturation to ensure adequate tissue perfusion and to allow the real-time optimization of clinical measures to balance the circulations. If the ductus arteriosus is widely patent, prostaglandin E1 is initiated at 0.01 mcg/kg/minute. The most common side effect is apnea and is often managed with intravenous caffeine, minimizing the need for preoperative intubation. Pulse oximetry is typically in the low to mid-90s for the relatively asymptomatic neonate with HLHS. Initial palliation most commonly occurs during the first week of life.

Operative management
Surgical management of HLHS involves three operations. The first and most significant is the Norwood procedure, which entails an atrial septectomy, aortic arch reconstruction with combination of the pulmonary artery and the hypoplastic aorta as a single outflow tract, and creation of a source of pulmonary blood flow either via a right ventricle to pulmonary artery conduit (Sano shunt) or via a shunt from the pulmonary veins.
Perioperative management
The patient with HLHS faces similar physiologic challenges prior to surgery, during the early postoperative period and through the interstage period until the second-stage palliation. The single right ventricle inefficiently pumps blood in parallel to the pulmonary and systemic vascular beds. Given the low pulmonary vascular resistance, systemic blood flow remains at risk regardless of optimal shunt size and placement. At the time of the second operation, the child is no longer shunt-dependent for pulmonary blood flow, and the heart pumps blood to the systemic vascular bed alone, leading to more efficient circulation.

To attenuate the expected increases in systemic vascular resistance at the time of the Norwood operation, we routinely use pharmacologic alpha blockade. This tactic, along with perioperative monitoring of venous oximetry and multisite near-infrared spectroscopy of oxygen delivery to the brain and kidney region, was pioneered at Children’s Hospital of Wisconsin and has been widely adopted across the globe. These strategic improvements that target optimizing oxygen delivery have paralleled improved survival and been linked to improved neurologic outcomes.

Home monitoring between Norwood operation and Stage 2 palliation
The average hospital stay after the Norwood operation is five weeks. Due to the persistence of high-risk, vulnerable circulation at discharge, vigilant monitoring is important to incorporate strategies to facilitate their development into routine clinical surveillance and care.

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