27 Hypoplastic Left Heart Syndrome

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Hypoplastic left heart syndrome (HLHS) describes varying degrees of underdevelopment of the left-sided heart structures that cannot independently support the systemic circulation (Figure 27-1). It is most commonly classified based on whether the aortic and mitral valves are atretic or stenotic into 4 types: aortic atresia/mitral atresia, aortic stenosis/mitral stenosis, aortic atresia/mitral stenosis, and aortic stenosis/mitral atresia. The most common variant of HLHS is aortic atresia, which results in significant hypoplasia of the ascending aorta and aortic arch. Patients with mitral stenosis/aortic atresia may have associated coronary fistulas, akin to the development of fistulas in patients with pulmonary atresia and intact ventricular septum (see Chapter 18).

Even though HLHS accounts for only 1-4% of all CHD, it causes one-fourth of all cardiac deaths in the first week of life and ~15% of deaths from CHD in the first month of life. There are no known genetic abnormalities linked to HLHS but it occurs more commonly in males. Most patients are born at term and rarely have other noncardiac anomalies. It is unusual to have structural brain malformations but approximately 20-25% of patients will have some preoperative MRI evidence of brain injury indicating varying degrees of immaturity of the brain.

Pathophysiology and Clinical Presentation

In HLHS, the LV is not functional, so the pulmonary venous return must be routed to the RA through a stretched PFO or an ASD. Systemic and pulmonary venous return mix in the right side of the heart. The RV then provides cardiac output to both the systemic and pulmonary circulations in a parallel fashion. Blood flows from the RV to the main PA, which provides flow to the lungs, and through the PDA to the descending aorta, brachiocephalic vessels, and coronary arteries (particularly in the case of aortic atresia). Ductal patency and an unrestrictive interatrial communication are thus crucial for survival in these neonates.

A majority of patients with HLHS are now diagnosed prenatally. This allows adequate family counseling, arranging for delivery in a tertiary care center, and immediate administration of PGE after birth to maintain ductal patency. At birth, most neonates have a balanced circulation. On exam, they tend to be warm and well perfused, but with some degree of cyanosis. On auscultation, they will have a single S_2 sound. Neonates with a restrictive interatrial communication will develop intense cyanosis/hypoxemia and respiratory distress from pulmonary venous congestion that is unresponsive to conventional medical management. If ductal patency is not maintained (e.g., patients with no prenatal diagnosis of HLHS), patients will develop poor systemic perfusion with signs of cardiogenic shock including lethargy, cool extremities, diminished pulses, hypotension, respiratory distress, and metabolic acidosis with end-organ ischemia.

The relationship between the amount of blood flow to the lungs (Qp) and the systemic circulation (Qs) depends on a delicate balance between PVR and SVR. After birth, as the PVR drops, an eventual imbalance in the parallel circulation will result in an increase

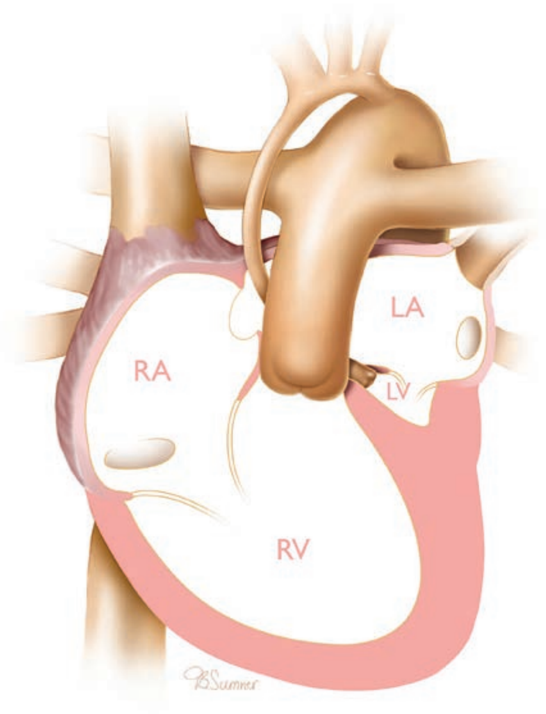

Figure 27-1. Anatomy of HLHS with a hypoplastic LV, a stenotic or atretic mitral valve, and a stenotic or atretic aortic valve. The ascending aorta is usually significantly hypoplastic and there are varying degrees of aortic arch hypoplasia. A large PDA and an unrestrictive ASD are important for survival.

in pulmonary blood flow through the PDA. Over the span of a few days, patients will start developing pulmonary overcirculation (Qp>Qs), leading to the development of CHF symptoms (i.e., tachypnea, diaphoresis, failure to thrive, pulmonary congestion) and reduced systemic circulation.

Diagnosis

- **ECG.** Classically shows RA enlargement and RV hypertrophy.
- **CXR.** Can reveal cardiomegaly and as the patient becomes overcirculated, it will show signs of pulmonary congestion.
- **Echocardiography.** Mainstay of diagnosis. It is important to show the anatomic details, interatrial communication, ductal patency, RV function, degree of TR, and pulmonary venous drainage.
- **CTA and cardiac catheterization.** Rarely used. However, in patients with mitral stenosis and aortic atresia, CTA and/or catheterization may be indicated to rule out the presence of significant coronary fistulas.

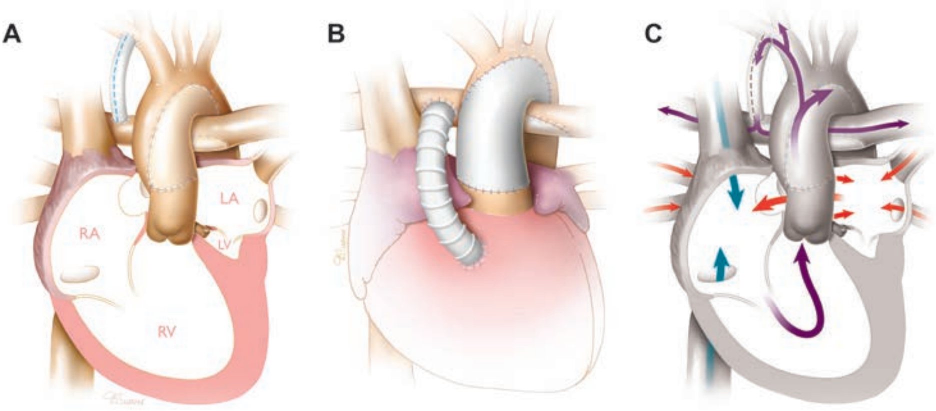

Figure 27-2. Norwood procedure. A) Norwood procedure with an mBTTS from the right subclavian artery to the right PA. B) Norwood procedure with an RV-PA conduit brought to the right of the aorta (Brawn modification). C) Hemodynamic flows after a Norwood procedure.

Preoperative Management

The primary goal of the management of the HLHS patient is to optimize systemic oxygen delivery and organ perfusion.

Immediately after delivery, arterial and venous access are obtained (UAC and UVC) in the delivery room. A PGE infusion (0.01-0.1 mcg/kg/min) is started to maintain systemic perfusion. An echocardiogram is necessary to rule out the presence of a restrictive interatrial communication. If there is significant restriction, the patient may need to undergo a balloon atrial septostomy (BAS). These patients usually need to wait for several days after the BAS to allow for resolution of organ dysfunction (from severe hypoxia) prior to any surgical palliation. Patients with mild restriction at the atrial level and no clinical embarrassment are better left alone as a BAS can otherwise lead to more overcirculation.

The preoperative management of the patient entails optimizing the balance between Qp and Qs. An excess of one will lead to compromise of the other. A few days after birth and while on PGE, the patient will develop pulmonary overcirculation with an increase in SaO_2 and pulmonary congestion. If left untreated, it can lead to systemic hypoperfusion, resulting in coronary ischemia and end-organ dysfunction. This may lead to mesenteric ischemia (necrotizing enterocolitis), acute kidney injury, and cerebral hypoxic-ischemic injury. The medical management goals thus include limiting Qp. This is partly achieved by avoiding pulmonary vasodilators like oxygen or respiratory alkalosis. Noninvasive ventilation to increase PVR can also be used. Diuretics can be used to decrease pulmonary congestion as the resulting tachypnea may lead to respiratory alkalosis. Careful use of systemic vasodilators (such as sodium nitroprusside infusion or milrinone) can also be utilized to reduce Qp and improve Qs. Eventually, if not taken for surgical palliation, these patients may become unstable and require intubation and mechanical ventilation.

Indications for intubation include apnea or severe respiratory distress, significant metabolic acidosis, significant pulmonary overcirculation, or end-organ dysfunction such as myocardial dysfunction. The ventilation management should target blood gases to limit pulmonary blood flow: $PaCO_2$ 35-45 mmHg, pH 7.35-7.40, FiO_2 21%, PEEP 4-5. The goal SaO_2 should be 75-85% (since this indicates close to a 1:1 Qp:Qs with this physiology), and the hemoglobin should be maintained at 14-16 g/dL.
At TCH, all neonates requiring cardiac surgical intervention undergo a brain MRI prior to surgery (see Chapter 52).

Anesthetic Considerations

The principles of intraoperative management of these patients remain the same – to maintain "balanced" systemic and pulmonary circulations. Standard anesthetic and cardiac monitors, NIRS, and transcranial Doppler (TCD) are routinely used. TEE is typically not used.

Intravenous induction is performed using synthetic opioids (fentanyl). Inhaled anesthetics are kept to a minimum or avoided prior to initiating CPB since they can significantly affect the patient's hemodynamics. The preferred route of intubation is nasotracheal. Immediately after intubation, the FiO_2 is reduced to 21%, if possible, to avoid overcirculation. The ventilator management strategy should be the same as mentioned above, with target $PaCO_2$ 35-45 mmHg, pH 7.35-7.40, FiO_2 21%, and PEEP 4-5 cmH_2O. The goal SaO_2 should be 75-85%, although it tends to be difficult to achieve due to the degree of overcirculation. Blended nitrogen, which was previously used on these patients, is not used anymore at TCH due to safety reasons.

A right-radial arterial line is useful with titration of CPB flows once antegrade cerebral perfusion (ACP) is initiated. A femoral arterial line or UAC is generally also placed to allow for monitoring of distal perfusion on CPB and adequate reads on the post-CPB period, since the radial artery is prone to spasm. In addition, if a right modified Blalock-Taussig-Thomas shunt (mBTTS) shunt is placed on the right subclavian artery, the reads from the arterial line may be falsely lower due to runoff into the pulmonary circulation. A femoral central line is routinely placed for CVP monitoring and infusion of medications during and after the procedure. The patient may require inotropic or vasopressor support prior to initiating CPB in order to maintain stable hemodynamics. Having the surgeon snare the right PA during initial dissection can also improve hemodynamics by reducing Qp. An initial dose of 100 Units/kg of heparin is administered prior to suturing the graft on the innominate/subclavian artery for ACP, and an additional 400 Units/kg of heparin are administered prior to initiating CPB.

During CPB, the patient is cooled down to 18 °C and vasodilators (e.g., phentolamine – an alpha-receptor blocking agent) are used to allow maximum organ perfusion while cooling. Baseline TCD measurements (measured through the anterior fontanel) and bilateral NIRS are established prior to ACP. CPB flows on ACP are titrated based on right-radial MAP, TCD, and NIRS.

Patients will require inotropic support to separate from CPB. Support usually includes an epinephrine infusion and a calcium-chloride infusion. Nitroprusside is used to counteract elevated BP. Platelets and cryoprecipitate are used to help stop bleeding after administration of protamine. The same principles in balancing Qp and Qs are used in the immediate post-CPB and postoperative periods.

Surgical Palliation

The diagnosis of HLHS is on itself an indication for surgical intervention. The surgical paradigm of HLHS has evolved significantly over the last few decades. The goal is to achieve a Fontan circulation through a staged approach that involves 3 operations: the Norwood procedure (performed at the newborn stage), the bidirectional Glenn (performed at 4-6 months of age), and the completion Fontan (usually performed between 3 and 5 years of age). For details of the bidirectional Glenn and the Fontan completion, please see Chapter 39. Long-term effects and management of the Fontan circulation can be found on Chapter 47.

The goals of the Norwood procedure (Figure 27-2) are: 1) to create an unobstructed outflow to the systemic circulation, 2) to allow unimpeded drainage of the pulmonary circulation, 3) to provide a stable source of pulmonary blood flow, and 4) to provide a reliable source of coronary blood flow. These goals are achieved by amalgamating the ascending aorta and the proximal PA (Damus-Kaye-Stansel [DKS] anastomosis), reconstructing the aortic arch, performing an atrial septectomy, and creating an mBTTS or placing an RV to PA conduit (Sano modification). Both mBTTS and RV-PA conduits are used at TCH. Patients that undergo an RV-PA conduit tend to have better hemodynamics postoperatively due to the lack of diastolic runoff seen in patients with mBTTS. Diastolic runoff causes the diastolic BP to be lower, potentially leading to coronary ischemia. On the contrary, it is more common for patients with RV-PA conduits to develop stenoses of the PA branches, potentially requiring intervention.

The Norwood procedure is performed via a median sternotomy. Due to the sometimes diminutive size of the ascending aorta, it is customary to mark with fine sutures the location where the ascending aorta kisses with the proximal PA to later facilitate the DKS anastomosis. It is sometimes useful to partially snare the right PA with a fine tourniquet at this time in order to improve hemodynamics by reducing overcirculation (the FiO_2 is increased as needed). The PDA and the brachiocephalic vessels are dissected and a 3.5 mm Gore-Tex® graft is sutured to the distal innominate artery/proximal right subclavian artery. If an mBTTS is expected, an effort is made to place the graft as far distal as possible on the subclavian artery since this graft would serve as the shunt (see Chapter 38). Particular care is taken not to injure the right recurrent laryngeal nerve that travels around the subclavian artery since temporary dysfunction of the left recurrent laryngeal nerve is not uncommon due to the dissection of the aortic arch and descending aorta.

CPB is instituted via the innominate/subclavian graft and a single venous cannula in the RA. The PDA is ligated with a 5-0 Prolene pursestring. The patient is cooled down to 18 °C. While cooling, the aortic arch, brachiocephalic vessels, PDA, and descending aorta are completely dissected. The PA may be divided at this time and the distal orifice closed, or partially closed, with or without a patch, depending on the plans for sourcing of pulmonary blood flow. It is important to dissect well the PA branches, in particular if an RV-to-PA conduit is expected to be placed. At this time, a decellularized homograft patch is prepared.

Once the goal temperature is reached, the head is packed on ice, the brachiocephalic vessels and descending aorta are controlled with fine tourniquets and CPB flows are reduced and stopped, thus initiating a short period of deep hypothermic circulatory arrest. Cardioplegia is administered through a sideport on the arterial limb of the circuit, therefore administering the cardioplegia through the innominate/subclavian graft and into the dimininutive ascending aorta. Future cardioplegia doses (every 20 minutes throughout the cross-clamp period) will be administered directly once the ascending aorta is open. After the initial cardioplegia dose is administered, the venous cannula is removed from the RA and working through the cannulation site, an atrial septectomy is performed. The venous cannula is replaced and a period of ACP is initiated through the innominate/subclavian artery graft. Flows are titrated using a combination of transcranial Doppler, NIRS, and right-radial arterial line pressure monitoring, and are usually between 30% and 40% of full CPB flows (see Chapter 6).

The PDA is divided and the aorta is opened longitudinally from the marking stitch on the proximal ascending aorta all the way to the descending aorta, past the insertion of the ductus arteriosus. The isthmus is divided and all ductal tissue is excised unless the left subclavian artery originates from the ductus arteriosus, in which case a small amount of posterior wall may need to be left in place. A posterior half-circumference anastomosis is performed between the descending aorta and the isthmus, the ascending aorta and the PA are anastomosed with fine sutures, and the remaining of the ascending aorta/aortic arch are reconstructed with the previously prepared homograft patch. After the aorta is reconstructed, all tourniquets around the brachiocephalic vessels are released, full-flow CPB is reinstituted, and the patient is rewarmed.

If an *RV-to-PA conduit* will be placed, a ringed Gore-Tex® graft is cut flush with one of the rings and "dunked" into the RV prior to finishing the aortic reconstruction. Usually, 2 rings are completely dunked into the RV and the third one is left flush outside of the RV. The graft is secured to the RV surface with a pursestring and a few interrupted stitches. In general, a 5 mm ringed graft is used for patients <3 kg and a 6 mm graft for those >3 kg. The graft may be brought to the left of the aorta (conventional Sano) or to the right side (Brawn modification). If an *mBTTS* will be used, the arterial limb of the circuit is changed to a cannula inserted into the reconstructed aorta, the previously placed graft on the subclavian artery is trimmed, and an anastomosis is performed between the graft and the right PA.

A peritoneal dialysis catheter is routinely placed on all patients undergoing the Norwood procedure and temporary atrial pacing wires are also usually placed. If the patient's hemodynamics and oxygen saturations are adequate, there is no bleeding, and there are no other concerns, the chest is routinely closed.

Other Management Strategies

There are several risk factors that significantly increase the risk of performing a Norwood operation. Some of these factors include prematurity, low-birth weight (<2.5 kg),

Recent TCH Norwood experience (2017-2018)

Number of Norwood procedures: 57 (49 primary, 8 after prior biventricular repair or hybrid)

Patients discharged home between first and second stages: 60%

Survival to second stage (bidirectional Glenn): 96%

1-year overall survival: 93%

1-year transplant-free survival: 89%

RV dysfunction, and significant TR. Even though a Norwood procedure is almost always used at TCH for palliation of HLHS, some high-risk patients may benefit from alternative strategies on a case-by-case basis.

The *hybrid approach* to HLHS has been used in some programs as an alternative to the Norwood operation. In general, this approach is not favored at TCH except in conditions where the risk of a Norwood operation may be prohibitive. The goals of the hybrid approach are the same as those of the Norwood operation (unobstructed systemic outflow, unimpeded pulmonary venous return, a stable source of pulmonary blood flow, and a reliable source of coronary blood flow) but achieved in a different way. The hybrid procedure consists of placement of bilateral PA bands, a BAS, and either placement of a PDA stent or continuation of PGE to maintain ductal patency ("chemical" hybrid). Patients with no prograde flow across the aortic valve that undergo placement of a PDA stent need to be closely monitored for development of a "retrograde arch malperfusion", i.e., obstruction of the aortic arch at the level of the stent obstructing retrograde flow into the arch, brachiocephalic vessels, and coronary arteries.

Listing for *neonatal heart transplantation* is another strategy that can be used on high-risk patients with HLHS. However, due to the overall shortage of organ donors, this strategy is fraught with risk and thus reserved for very particular circumstances. Due to the risk of overcirculation, it is not unusual to proceed with placement of bilateral PA bands or a hybrid approach as a bridge to transplantation.

Comfort care is a strategy that may be used by some families in cases in which the risk of palliation is prohibitively high. However, this is a strategy rarely used nowadays.

Postoperative Management

The postoperative management of patients after the Norwood procedure follows some of the same guidelines described for the preoperative and intraoperative management. Overall, the goal is to maintain adequate system oxygen delivery and organ perfusion by allowing a careful balance of pulmonary and systemic circulations (Qp:Qs).

Hemodynamics and oxygen saturations are carefully monitored. Patients usually arrive to the CICU with a combination of inotropes including low-dose epinephrine and calcium. Milrinone may be started in the OR or is sometimes added in the CICU. Patients with an RV-to-PA conduit tend to have better diastolic BP than those with an mBTTS due to the lack of diastolic runoff. As such, some patients with mBTTS may require addition of vasopressin in order to improve the diastolic BP and thus coronary perfusion. See Chapter 38 for different clinical postoperative scenarios and recommended interventions in postoperative patients with a shunted circulation, scenarios that are directly applicable to patients after a Norwood operation.

Mechanical ventilation is slowly weaned with the goal of extubating patients within the first 24-48 hours. However, some patients, especially those that were intubated preoperatively, may require a longer intubation. Peritoneal dialysis is routinely started on the day of surgery and continued until the patient mobilizes fluids and urine output increases, usually within the first few days. Prophylactic heparin at 6 U/kg/hr is started 6 hours after surgery if there is no bleeding. It is then converted to aspirin once the patient starts taking PO.

Enteral feeds via nasogastric tube are slowly started if the patient is expected to be intubated for a longer period of time. Otherwise, they are slowly advanced once the patient is extubated. It is not unusual for patients after a Norwood procedure to have some degree of feeding intolerance, likely due to the marginal gut circulation. This mandates slow progression of feeds as per protocol (see Chapter 58). In general, feedings are not fortified beyond 24 kcal/oz in patients after a Norwood procedure.

For details regarding requirements for transfer from CICU to the ward, interstage management, discharge planning, and home monitoring, see Chapter 39.