Understanding the Hybrid Stage I Approach for Hypoplastic Left Heart Syndrome

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Hybrid stage I palliation combines cardiothoracic surgery and interventional transcatheter procedures for treatment of hypoplastic left heart syndrome. The approach is an alternative to the Norwood procedure, the traditional first stage of surgical palliation. Hybrid stage I palliation involves placing bilateral branch pulmonary artery bands and a patent ductus arteriosus stent through a median sternotomy, performed without cardiopulmonary bypass. The purpose of the bands is to control blood flow to the lungs and protect the pulmonary bed while the stent sustains systemic cardiac output. A balloon atrial septostomy is performed to create an atrial septal defect for unobstructed blood flow from the left atrium to the right atrium. The second stage of palliative surgery is the comprehensive stage II, which incorporates removal of the stent and pulmonary artery bands, atrial septectomy, anastomosis of the diminutive ascending aorta to the main pulmonary artery, aortic arch augmentation, and bidirectional cavopulmonary anastomosis. The traditional Fontan procedure completes the series of palliation. (Critical Care Nurse. 2016;36[5]:48-55)

The hybrid stage I procedure (Figure 1) is an alternative, lower risk palliation for newborns with hypoplastic left heart syndrome (HLHS) and has been previously described.¹ The hybrid procedure, unlike the traditional Norwood procedure or Norwood variation, avoids cardiopulmonary bypass, circulatory arrest, and associated surgical risks early in the neonatal period. Use of the hybrid procedure transfers the major open heart surgery, including the risks associated with cardiopulmonary bypass, to early infancy, usually between 4 to 6 months of age; the premise is that cardiopulmonary bypass will be better tolerated at that age.

CE 1.0 hour

This article has been designated for CE contact hour(s). The evaluation tests your knowledge of the following objectives:
1. Identify the surgical and interventional techniques that compose the hybrid stage I procedure
2. List the advantages of the hybrid stage I procedure
3. Describe monitoring required of infants with hypoplastic left heart syndrome during the interstage period

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Anatomy and Physiology

The diagnosis of HLHS may include aortic atresia with mitral atresia, aortic atresia with mitral stenosis, or aortic stenosis with mitral stenosis. Cardiac output is primarily, if not completely, dependent on the patent ductus arteriosus (PDA). The right ventricle is the systemic pumping chamber; it pumps blood across the pulmonary valve to the main pulmonary artery, through the right and left pulmonary arteries to both lungs, and across the PDA to the systemic circulation. The ductus arteriosus is kept patent by intravenous infusion of prostaglandin E1. During the first several weeks of life, the pulmonary artery resistance decreases, resulting in pulmonary overcirculation.

During retrograde perfusion of the aortic arch, oxygenated blood flows from the PDA backward to the aortic arch, carotid arteries, ascending aorta, and coronary arteries. This retrograde flow is the only source of oxygenated blood to the heart and to the brain for neonates with HLHS who have aortic atresia rather than aortic stenosis. Underperfused myocardium can result in ischemia and changes in electrocardiographic findings.

The atrial septal defect (ASD) allows unobstructed blood flow to return from the lungs to the left atrium and across the ASD. If the defect becomes smaller and more restrictive, the pressure difference increases in the left atrium and in turn may create pulmonary hypertension and signs of congestive heart failure. The first stage of palliation, whether the Norwood procedure or the hybrid procedure, establishes dependable systemic blood flow, limits pulmonary overcirculation, and provides unobstructed blood flow across the atrial septum.

Technique

Hybrid stage I palliation combines cardiothoracic surgical and interventional transcatheter techniques for treating HLHS during the same operative procedure and operative setting. This management approach, typically performed within the first week of life, involves placing pulmonary artery bands (PABs) on both branches of the artery and a PDA stent through a median sternotomy on the beating heart, without cardiopulmonary bypass. A balloon atrial septostomy is performed, either during this procedure or at a later date, to create an adequate-sized ASD or opening between the left and right atria, to allow unobstructed blood flow returning from the lungs to the left atrium to shunt across the ASD to the right atrium.

After a median sternotomy, the cardiothoracic surgeon places a 1- to 2-mm–wide synthetic vascular graft (Gore-Tex band; W. L. Gore & Associates Inc), approximately 3.0 to 3.5 mm in diameter, around the proximal left and right pulmonary arteries. This step is followed by placement of either a self-expandable or a balloon-expandable stent in the PDA by the interventional cardiology team. The stent is placed through a sidearm sheath that is placed approximately 2 mm into the main pulmonary artery after direct puncture and secured with a purse string suture. A small amount of contrast material is injected through the sidearm of the sheath to delineate the anatomy of the PDA, descending aorta, and retrograde aortic arch. Appropriate measurements of the PDA at

Figure 1 Drawing of hypoplastic left heart syndrome shows right and left pulmonary artery bands, patent ductus arteriosus (PDA) stent, and balloon atrial septostomy of the hybrid stage I procedure. Copyright Nationwide Children’s Hospital, reprinted with permission.
the pulmonary entrance, mid ductus, and distal ductus, along with ductal length, are obtained to determine the size and type of stent. If any evidence of stenosis is detected, a balloon-expandable stent is used. If no stenosis is present, an alternative is to use a self-expanding stent. The key is to ensure that the entire length of the ductus is covered by the stent to prevent the ductal tissue from closing and becoming stenotic. Once the PDA stent is in place, the infusion of prostaglandin E1 can be discontinued. An alternative to placing a stent in the PDA is to continue infusion of prostaglandins after placement of the PABs as a bridge to the Norwood procedure. Creating an adequate-sized ASD or larger hole between the right and left atria, usually via balloon atrial septostomy, finalizes stage I palliation. Occasionally, a stent is required to ensure an adequate ASD, particularly if the septum is thick or balloon atrial septostomy is not successful.

**Postoperative Course**

The immediate postoperative course in a neonate who has had a Norwood procedure differs markedly from the postoperative care after a hybrid procedure. Compared with a Norwood procedure, the hybrid procedure is associated with shorter time to extubation, fewer blood transfusions, initiation of enteral feedings, and shorter stays in the intensive care unit and hospital. However, whether or not oxygenation and perfusion to the brain after a hybrid procedure are adequate, because of the persistent retrograde aortic blood flow, remains to be answered. Galantowicz et al reported that 52% of patients were extubated within 24 hours. Inotropic support was not required in any patient, and 79% of patients were feeding within 24 hours. No patient required extracorporeal membrane oxygenation or delayed sternal closure. Hospital survival to discharge was 97.5%, mean length stay of in the cardiothoracic intensive care unit was 4.5 days, and mean postoperative hospital length of stay was 13 days. Interstage medications after placement of the PDA stent may include aspirin, furosemide, digoxin, or enalapril.

Much of the postoperative nursing care focuses on monitoring vital signs and oxygen saturation and assessing for signs of respiratory distress. Arterial blood gases and electrolytes and lactate levels are monitored in the initial postoperative period; the results inform postoperative management. Other aspects of nursing care after a hybrid stage I palliation are pain management, wound care, and growth and nutrition (parenteral and enteral). Nurses should observe patients for signs of feeding intolerance (ie, decreased oral intake), increased work of breathing with retractions, tachypnea, diaphoresis, and lack of weight gain. Any changes should be reported to the medical team.

**Growth and Nutrition**

The focus of the National Pediatric Cardiology Quality Improvement Collaborative of the Joint Council on Congenital Heart Disease has been on decreasing interstage mortality. Although preoperative enteral feedings have been controversial, the collaborative feeding group has established algorithms and feeding guidelines for single-ventricle patients. Total parenteral nutrition is recommended, with advancement to full caloric intake of 90 to 100 kcal/d. Amino acids should be started at 1.5 to 3 g/kg per day and increased by 1 to 1.5 g/kg per day to a maximum of 3 g/kg per day. Lipids should be started at 1 to 3 g/kg per day and increased by 0.5 to 1 g/kg per day to a maximum of 3 g/kg per day. Adequate caloric goals can be met during interstage monitoring, regardless of the feeding technique. Initiation of enteral feeding is strongly recommended if the patient’s hemodynamic status is stable, regardless of use of an umbilical catheter or prostaglandin E1 infusion. A nasogastric feeding tube may be used; however, the collaborative group has made no specific recommendation on this practice. The goal is to eventually reach a feeding volume of 120 to 140 mL/kg per day and 120 to 150 kcal/kg per day.

**Advantages and Disadvantages**

The major advantages of the hybrid stage I procedure include avoiding cardiopulmonary bypass, circulatory arrest, and the associated risks of open heart surgery in the neonatal period. Other advantages include early extubation, no need for inotropic support, no blood transfusion, decreased intensive care length of stay, and early feedings. Hybrid stage I palliation can be performed on premature babies weighing less than 2.5 kg. Neonates weighing less than 2 kg have successfully undergone hybrid stage I palliation. However, these extremely
premature babies remain at risk for other comorbid conditions associated with prematurity, such as necrotizing enterocolitis. Schranz et al\textsuperscript{14} reported an 84\% survival at 1 year for children with HLHS who underwent the hybrid procedure, with a 15-year survival rate of 77\%. Risk factors of low birth weight or aortic atresia did not affect the 15-year outcome data.\textsuperscript{14}

Some institutions reserve the hybrid stage I procedure for extremely high-risk patients (ie, infants who weigh less than 2.5 kg or have other comorbid conditions such as extreme prematurity) who would otherwise be extremely high risk for mortality with the Norwood procedure.\textsuperscript{15-18} Doing so may be considered an advantage in terms of weight, or a disadvantage because high-risk patients may have the highest risk for mortality. Other disadvantages include the need for close monitoring, possibly every 2 weeks. This monitoring need may be an added burden for the patient’s family, particularly if they do not live locally and must travel for outpatient clinic appointments. Families may spend hours at each outpatient visit for complete assessment by an interprofessional team.

**Discharge and Home Monitoring**

The nursing team should help coordinate provision of appropriate therapies, such as occupational, physical, and speech therapies. Infants may need assistance with growth and development, as well as with deficits attributed to hospitalization. Nurses also play an important role in educating and supporting patients’ families in at-home care of the infant and this complex anatomical lesion. Before an infant is discharged from the hospital, parents and caregivers may need education and instructions on tube feedings, signs of respiratory distress, and cardiopulmonary resuscitation. Members of the nursing team help coordinate discharge planning and follow-up, alerting community emergency units and the pediatrician of the infant’s status and planned discharge. Nurses are key in the coordination of care to help achieve the best outcomes.

Home monitoring for patients with a single ventricle has been reported.\textsuperscript{19-24} Infants with HLHS who have had hybrid stage I palliation need to be closely monitored after hospital discharge. Incorporating a home-monitoring program after either hybrid or Norwood stage I palliation is recommended. Parents or caregivers should monitor the infant’s daily caloric intake by recording daily formula intake, daily weight, and daily oxygen saturation (via pulse oximetry) to evaluate for potential complications.\textsuperscript{19-24} Such monitoring may lead to a marked decrease in mortality.\textsuperscript{25} Data are generally reported to the home-monitoring nurse on a weekly basis. Parents are instructed when to call if a deviation from the established criteria occurs to determine whether further clinical assessment or possible hospitalization is warranted. At Nationwide Children’s Hospital, Columbus, Ohio, outpatient visits for patients with a single ventricle include a careful feeding history, review of home-monitoring data, physical examination, an electrocardiogram, and an echocardiogram at each visit. Additionally, patients are evaluated by a nutritionist and by an occupational or a physical therapist or both.

For HLHS patients who have feeding difficulty or increased cyanosis or whose findings on surveillance echocardiography are a concern, closer observation is required. Physical examination in the outpatient cardiology clinic should include measurement of blood pressure in the upper and lower extremities (both arms and at least 1 leg), along with examination of right brachial and femoral pulses to check for signs of retrograde aortic arch obstruction or evidence of coarctation of the descending aorta. Both echocardiograms and electrocardiograms should be obtained and should be compared with previous findings.

During the interstage period, growth and nutrition must be assessed continually, and nurses play a key role in the assessment. Monitoring should include daily weight and total 24-hour volume and caloric intake of feedings; the goal is a weight gain of 20 to 30 g/d.\textsuperscript{10,11} As part of the home-monitoring program, a lack of success in reaching these nutritional guidelines is indicated by weight loss greater than 30 g in 1 day, failure to gain 20 g in 3 days, and intake less than 100 mL/kg per day.\textsuperscript{10,23,24} Such changes may prompt an assessment by the cardiology team and possible hospital admission.

A registered dietician is a vital part of the interprofessional management team for patients with HLHS and should be involved in all nutritional assessments and issues.\textsuperscript{10,23,24} Miller-Tate et al\textsuperscript{23} reported a mean interstage weight gain of 16.85 g/d (SD, 5.94 g/d) in HLHS
patients who underwent the hybrid stage I procedure at a mean of 6.12 months (SD, 1.37 months) of age. The mean weight z score before stage II was -2.25 (SD, 1.28). Patients who received home monitoring had significantly higher weight z scores (mean, -1.67; SD, 0.98) compared with patients without home monitoring (mean, -2.82; SD 1.28). These interstage weight gains are comparable to gains after the Norwood procedure. Regardless of the type of palliation, home-monitoring programs and feeding protocols have significantly improved outcomes in growth and nutrition.

Nurse-managed home-monitoring programs are helpful in providing support to patients’ families while monitoring the patients’ nutrition, weight gain, oxygen saturations, and respiratory status. Caring for an infant with HLHS at home and monitoring the infant’s growth and respiratory status can be quite burdensome for the infant’s family members and are often highly stressful. Much support is needed for families of infants with HLHS, from social service personnel, nurses, registered dieticians, occupational and speech therapists, physical therapists, and the team of cardiologists and the cardiothoracic surgeon. Meetings with parents and the home-monitoring team via the computer applications FaceTime or Skype may help alleviate some anxiety and give the care team a better assessment of the infant’s status.

Concerns During the Interstage Period

The interstage period between hybrid stage I and surgical comprehensive stage II procedures is associated with many risk factors. Infants in this stage reach their nadir in hemoglobin levels, and their oxygen-carrying capacity may be at the lowest. Major limitations of the hybrid procedure are the persistent cerebral and myocardial perfusion during the first several months of life and the potential risk for retrograde aortic arch obstruction resulting in decreased cerebral and myocardial oxygenated blood flow. Decreased cerebral and myocardial perfusion may be devastating if not monitored closely and if appropriate interventions are not done in a timely fashion. Other problems may also arise during the interstage period, including an increased gradient across the ASD, intimal proliferation causing in-stent stenosis in the PDA stent, or recoarctation distal to the PDA stent. All of these require urgent intervention, such as creation of a larger ASD or possibly stent placement in the atrial septum, coaxial stent placement in the PDA stent, or stenting of the retrograde aortic arch. Any interstage weight gains are comparable to gains after the Norwood procedure. Regardless of the type of palliation, home-monitoring programs and feeding protocols have significantly improved outcomes in growth and nutrition.

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Indications for Cardiac Catheterization

In infants who have had the hybrid stage I procedure, echocardiographic indications of a decrease in qualitative right ventricular function or an increase in the degree of tricuspid regurgitation or both are often associated with obstruction and require close interim follow-up. Any electrocardiographic evidence of continued or marked decrease or new ST-T wave changes or an increase in retrograde aortic arch velocity may require hospital admission and consideration of cardiac catheterization. Any abnormal diastolic flow pattern may indicate an increase in distal pulmonary artery pressure and development of pulmonary vascular resistance. Overall, echocardiographic assessment is vitally important. However, echocardiography is limited for grading the degree of function, which is a subjective finding, or for assessing for in-stent stenosis due to intimal proliferation. The latter is not evident on echocardiograms. Evidence of acceleration of blood flow on Doppler imaging is indicative of stenosis, which may cause obstruction of blood flow. Often, a team discussion of a strategic plan is beneficial, particularly in terms of cardiac catheterization and intervention (eg, conversion to a Norwood stage I procedure or possibly an early stage II palliation).
Comprehensive Stage II Procedure

The second-stage reconstruction after hybrid stage I palliation is referred to as the comprehensive stage II procedure (Figure 2). This reconstruction is considered the open heart surgery for the staged palliation. Essentially, the surgery involves part of the traditional Norwood procedure, as well as the second stage palliation. The comprehensive stage II procedure incorporates removal of the PDA stent and PABs, excision of the atrial septum, anastomosis of the diminutive ascending aorta to the main pulmonary artery, augmentation of the aortic arch, and a cavopulmonary anastomosis. Therefore, this second-stage palliation involves prolonged duration of cardiopulmonary bypass. This prolonged duration must be carefully considered when the hybrid path is taken.

Timing of this second-stage procedure may be influenced by the retrograde aortic arch blood flow or recoarctation or both. Pulmonary artery pressures may also influence timing for a cavopulmonary anastomosis. High pulmonary artery pressures may result in increased cyanosis in anatomical regions with passive venous blood flow with the cavopulmonary anastomosis. The comprehensive stage II procedure is usually performed when the infant is 6 months old. However, the idea that cardiopulmonary bypass and the associated risks of a major open heart surgery are better tolerated at 4 to 6 months of age than during the neonatal period has not been proved. If an HLHS requires an early comprehensive stage II procedure, within the first few months after the hybrid stage I, a better alternative may be conversion to a traditional Norwood procedure. However, both conversion to a stage I Norwood procedure and an early comprehensive stage II procedure may be practical options, depending on the age and weight of the patient, particularly if the infant has systemic outflow obstruction.

Angiography performed before cardiopulmonary bypass is stopped and the chest is closed is recommended to evaluate the cavopulmonary connection and branch pulmonary artery flow. The angiography can be performed by using a portable C-arm for fluoroscopic imaging and inserting an angiographic catheter directly into the superior vena cava, temporarily stopping or decreasing blood flow in the cardiopulmonary bypass circuit, and using a power injector to deliver the contrast material for imaging. The results of angiography may indicate a need for immediate therapy, either surgical or transcatheter. The angiographic findings are also helpful during the postoperative management period and allow for possible plans for future early assessment or intervention via cardiac catheterization. An alternative option for obtaining an angiogram may be a surveillance cardiac catheterization, particularly if the patient continues to require ventilatory support in the postoperative period.

Fontan Procedure

The Fontan procedure is the third and final stage of reconstruction in patients with HLHS and is essentially the same operation for children born with the syndrome who have the traditional Norwood staged procedure. The Fontan procedure is the total cavopulmonary connection in which the inferior vena cava is now anastomosed to the pulmonary artery. In general, the risks are low for complications associated with the Fontan procedure; however, pulmonary artery pressures are an important variable.
Developmental Outcomes

Neurocognitive development and long-term outcomes in HLHS patients who were initially palliated by hybrid stage I and comprehensive stage II procedures are unknown at this time, and published reports on the topic are limited. In one study, mortality at 1 year of age in patients with HLHS and other univentricular heart defects who underwent hybrid stage I palliation (n = 9) did not differ significantly from that of patients who had the Norwood procedure (n = 11). Additionally, the psychomotor development index and mental development index scores were similar in the hybrid and Norwood groups, and significantly lower in both groups than in normal healthy 1-year-olds. Similar to findings in previous developmental studies in children who had a Norwood palliation, motor impairment was high.40 In another study,41 infants with HLHS who underwent hybrid stage I palliation and normal, healthy, age-matched controls were compared. The HLHS group scored lower than the control group in motor skills, language, and cognitive development as indicated by scores on the Bayley Scales of Infant and Toddler Development.42 However, only scores for motor skills were significantly lower in the HLHS group (P = .049).41 More research is needed to determine longer term outcomes.

Conclusion

Hybrid stage I palliation is an alternative to the Norwood procedure for treatment of infants with HLHS. In the neonatal period, compared with the Norwood procedure, the hybrid procedure has several benefits, such as avoiding use of cardiopulmonary bypass and the systemic effect of the inflammatory process. Initially after surgery, patients who have the hybrid procedure generally require little intensive care support, unless they have other comorbid conditions. Once the patient is discharged, the home-monitoring team becomes an integral part of the infant’s care. Frequent monitoring and echocardiographic imaging with a complete comprehensive evaluation at an interprofessional outpatient clinic every 1 to 2 weeks are common. Patients who have had the hybrid procedure are vulnerable during this interstage period. Inherent potential risks include development of PDA in-stent stenosis, retrograde aortic arch obstruction, possible recoarctation of the aorta at the distal PDA stent, and a restrictive atrial septum. If any of these risks actually occur, then surgical or transcatheter interventions, or even a conversion to a Norwood palliation, may be required. The comprehensive stage II procedure, usually performed after the hybrid stage I procedure when the patients are 4 to 6 months old, is the big open heart surgical palliation that involves a prolonged duration of cardiopulmonary bypass. Although not proved, the premise is that the patient may better tolerate cardiopulmonary bypass later in infancy than in the early neonatal period.

The hybrid approach may not be appropriate for all neonates born with HLHS or for all institutions. Some institutions reserve this approach for extremely high-risk neonates, whereas others routinely or exclusively use the hybrid stage I procedure.14 The hybrid procedure has been used as a high-risk salvage procedure in neonates who are in extremis after a late diagnosis.38,43,44 It can also be performed as a bridge to transplant38,45 or 2-ventricle repair4,38 or as a bridge to the Norwood procedure.38,46

For standard-risk HLHS infants, outcomes after hybrid stage I procedure are comparable to outcomes after a Norwood procedure.2,4,43 Information on longer term outcomes is needed to determine which cohort of babies born with HLHS may benefit most from the hybrid procedure.

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