Surgical Treatment for Hypoplastic Left Heart Syndrome

Once considered a uniformly fatal condition, the outlook for newborns with hypoplastic left heart syndrome has been dramatically improved with either a protocol of staged reconstruction or cardiac transplantation. Currently, a significant shortage of suitable donor hearts restricts the applicability of transplantation for most newborns. At the University of Michigan, we have adopted a policy of staged reconstruction for all patients with hypoplastic left heart syndrome, reserving transplantation only for those unsuitable for reconstructive techniques. Between January 1990 and September 1998, 303 patients underwent the Norwood operation for classic hypoplastic left heart syndrome with an overall hospital survival of 76%. Among patients considered at standard risk, survival was significantly higher (86%) than that for those patients with important risk factors (42%), p=0.0001. Adverse survival was most strongly associated with significant associated noncardiac congenital conditions (p=0.008) and severe preoperative obstruction to pulmonary venous return (p=0.03). Survival following second stage reconstruction with a hemi-Fontan or bidirectional Glenn procedure was 98%. The Fontan procedure has been completed in 117 of these patients with a hospital survival rate of 91%. Survival after the Fontan procedure improved significantly when the second stage of the reconstruction was completed with a hemi-Fontan procedure compared to a bidirectional Glenn, 98% vs 81%, p<.05. Among the patients considered at standard risk, actuarial survival was 70% at 5 years. The largest decrease in survival occurred in the first month of life and late deaths affected primarily those patients in the high risk group. Neurodevelopmental outcome studies demonstrated normal verbal and performance scores in the majority of patients. Among centers utilizing a protocol of transplantation, donor organ shortages have resulted in a mortality of approximately 25% while awaiting transplantation with 5 year survival rates for those actually receiving organs essentially equal to those for staged reconstruction. Staged reconstruction and transplantation have significantly improved the intermediate-term outlook for patients with hypoplastic left heart syndrome. Factors addressing improvements in early first stage survival following the Norwood would be expected to add significantly to an overall improved late outcome. Outcome following cardiac transplantation is limited by donor availability in addition to the late complications of infection, rejection, graft atherosclerosis, and lymphoproliferative disease.
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Index words: HLHS (hypoplastic left heart syndrome), Norwood operation, hemi-Fontan procedure, bidirectional Glenn procedure, cardiac transplantation

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Over the past decade, treatment strategies with transplantation or staged reconstruction for hypoplastic left heart syndrome (HLHS) have been employed with increasing success, significantly improving the outlook for newborns with this oth-

erwise lethal condition. Staged reconstruction for patients with HLHS is based on the knowledge that an effective circulation is possible in the absence of a pulmonary ventricle. The principle that pulmonary blood flow could be maintained when certain well-defined hemodynamic criteria were met has led to the application of the Fontan procedure to virtually all forms of single ventricle lesions.[1] The first stage in the reconstructive process, the Norwood procedure, must provide: (1) unobstructed systemic and coronary blood flow from the right ventricle, (2) unobstructed pulmonary venous return across the atrial septum, and (3) sufficient pulmonary blood flow without significant volume overload.[2-4] This leaves the pulmonary and systemic circulations in parallel and the right ventricle to perform the increased volume work of both circulations. The second stage procedure, the hemi-Fontan or bidirectional Glenn operation, results in removal of the ventricular volume overload imposed by the systemic shunt and the connection of the superior vena cava to the pulmonary arteries.[5,6] Augmenting the central pulmonary arteries, avoiding conduction disturbances, and constructing a potential connection for the inferior vena cava to the pulmonary arteries are considered important components of this procedure.[7,8] The hemi-Fontan procedure, currently preferred to the bidirectional Glenn at the University of Michigan, also allows for the correction of additional risk factors, including pulmonary artery distortion, and maintains a stable circulation free of right ventricular volume overload and pulmonary hypertension. During the last stage of reconstruction, the Fontan procedure, inferior vena caval return is channeled to the pulmonary arteries to complete the separation of the pulmonary and systemic circulations. Although the current techniques have resulted in substantial improvements in the quantity and quality of survival, efforts to refine each stage of the process continue to evolve with increasing follow-up and evaluation.[7,9,10]

Many institutions, initially discouraged by the poor results of palliative reconstructive surgery for hypoplastic left heart syndrome, abandoned this approach in favor of transplantation.[11,12] However, the lack of available donor hearts, late complications of cardiac transplantation, and the improvements noted with staged reconstruction have again encouraged an increasing number of centers in North America and elsewhere to re-evaluate this form of therapy. At the University of Michigan, we have continued to pursue a policy of offering staged reconstructive surgery for patients with HLHS.

Methods

Preoperative management and the Norwood procedure. The majority of newborns with HLHS are well palliated with an infusion of prostaglandin and reduction of the inspired oxygen concentration to avoid high systemic arterial saturations, but this condition cannot be sustained indefinitely and a progressive deterioration in pulmonary function characterized by increasing pulmonary edema will become evident with time. The presence of significant obstruction to pulmonary venous return, most commonly resulting from a restrictive atrial septal defect, will hasten this outcome. In general, when systemic arterial oxygen saturation is in excess of 80%, peripheral perfusion is well maintained, and the chest radiograph shows little or no pulmonary edema, most patients will remain stable and may undergo operation electively within a few days. More significant degrees of obstruction to pulmonary venous return, however, will result in severe and rapidly progressive pulmonary edema with systemic arterial oxygen saturations falling below 75–80%. Urgent intervention is often required before the pulmonary status deteriorates further. Percutaneous balloon septostomy, often requiring a bioptome to traverse the thickened atrial septum, has been effective in our institution. Operation can then be postponed for a few days to allow the pulmonary edema time to resolve. Optimal results following the Norwood procedure are obtained when pulmonary blood flow is restricted with the use of a small shunt.[13,14] Satisfactory postoperative oxygen saturations are increasingly difficult to maintain, however, when a small shunt is used in a patient with pulmonary edema. A larger shunt will be required which will then often result in pulmonary overcirculation after only a few hours following cardiopulmonary bypass as the pulmonary edema and elevated pul-

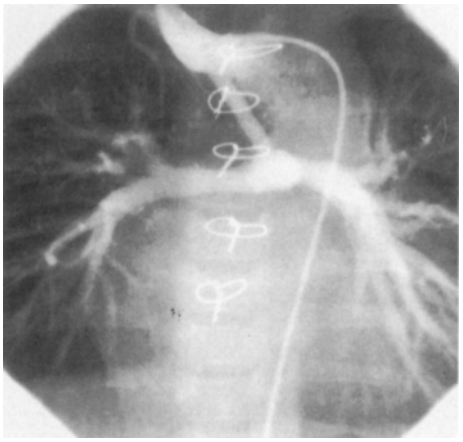

Fig. 1. Reconstruction following the Norwood procedure demonstrates the shunt which is positioned between the distal innominate or proximal subclavian artery and the central pulmonary arteries. Appropriate tailoring of the allograft patch is essential to avoid excessive dilatation of the ascending aorta and compression of the left pulmonary artery.
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monary vascular resistance resolves. A rapid deterioration in systemic perfusion associated with increasing pulmonary blood flow may result in hypotension, metabolic acidosis and death. A significant diastolic runoff of blood into the pulmonary circulation will also occur with a large shunt, further adding to low cardiac output from impaired coronary blood flow due to low diastolic blood pressure.[15]

Surgical technique. Cardiopulmonary bypass is established by cannulation of the proximal main pulmonary artery just distal to the pulmonary valve and the right atrial appendage. The branch right and left pulmonary arteries are occluded and systemic cooling is begun in preparation for circulatory arrest. The ascending, transverse and proximal descending segments of the aorta are mobilized and tourniquets are placed around each head vessel for subsequent occlusion. A cryopreserved pulmonary allograft is trimmed to fashion a patch that will serve to augment the aorta and allow anastomosis to the proximal main pulmonary trunk. Following the induction of circulatory arrest, the atrial septum is excised through the atrial appendage purse string or a short right atriotomy. The main pulmonary trunk is then divided at the bifurcation and the distal end closed with a synthetic patch of goretex. The ductal tissue is then excised completely and the resultant opening is extended approximately 10 mm further distally into the descending aorta. This incision is then extended proximally under the transverse arch and down the diminutive ascending aorta until the level of the previously divided main pulmonary trunk is reached. The ascending aorta is connected to the main pulmonary artery with interrupted sutures in the proximal corner near the coronary arteries and the remainder of the aorta is augmented with the allograft patch. A 3.5 mm goretex conduit is anastomosed from the subclavian or distal innominate artery to the central pulmonary artery during the rewarming (Fig. 1). A 4 mm conduit is occasionally used for patients weighing over approximately 4 kg, those with significantly increased extravascular lung water, or those who are beyond the first month of life. The distal end of the shunt is placed close to the divided end of the ductus rather than onto the right pulmonary artery itself.

The second stage: Hemi-Fontan procedure. At the University of Michigan, the hemi-Fontan procedure is preferred to the bidirectional Glenn because of the ability to more easily augment the central pulmonary arteries and to provide a large connection for the inferior vena cava at the subsequent Fontan procedure. Although increased in complexity, the hemi-Fontan procedure significantly simplifies the Fontan, at which time optimal hemodynamics and pulmonary function are the most essential to a good outcome. The hemi-Fontan procedure is generally performed between 4 and 6 months of age. Cardiac catheterization is routinely performed to assess: (1) right ventricular function, (2) tricuspid regurgitation, (3) residual arch or pulmonary venous (atrial septal) obstruction, (4) pulmonary artery anatomy, and (5) pulmonary vascular resistance. The number of patients with contraindications to proceeding with the second stage procedure has been surprisingly few. Absolute contraindications have included isolated poor right ventricular function and elevated pulmonary vascular resistance. However, when decreased right ventricular function exists in association with another correctable problem, typically

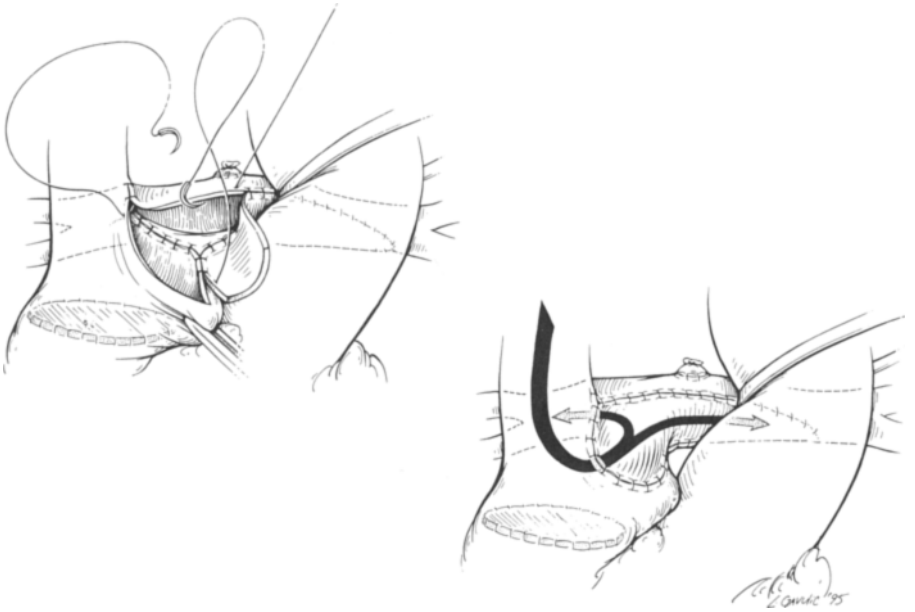

Fig. 2. The steps for the second stage reconstruction or the hemi-Fontan procedure. The central pulmonary arteries are opened anteriorly and an incision is made along the base of the right atrial appendage (top left). A patch is fashioned from a cryopreserved pulmonary allograft and used to augment the central pulmonary arteries (bottom right). An additional patch is placed within the right atrium to exclude the superior vena cava and the pulmonary arteries from the heart. Although not illustrated in this figure, the atrial incision does not cross the cavoatrial junction to avoid injuring the artery to the sinoatrial node.
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residual arch obstruction, excessive volume overload from a large shunt, or tricuspid valve regurgitation, we have proceeded with the hemi-Fontan with the expectation that ventricular function will improve. The belief that tricuspid valve regurgitation was secondary to volume overload and that it would improve after the second stage procedure has not proven to be the case, and we now perform tricuspid valvuloplasty at the hemi-Fontan procedure when regurgitation is more than mild.[16] Although placement of the shunt on the central pulmonary artery near the ductal insertion has resulted in more symmetric growth of the pulmonary arteries following the Norwood procedure, it has been common to find mild narrowing at the origin of one or both pulmonary arteries. The use of pulmonary allograft patches of smaller widths at the time of the Norwood procedure has also improved the arch reconstruction and reduced the incidence of compression of the left pulmonary artery.

Surgical techniques. Cannulation for cardiopulmonary bypass is accomplished through the distal ascending aorta, placing the arterial cannula through allograft patch tissue, and the venous cannula through the mid right atrial wall. The shunt is divided and the pulmonary arteries are mobilized widely. A patch of pulmonary allograft material is fashioned for augmentation of the pulmonary arteries. The superior vena cava is mobilized and the azygous vein is ligated. The central pulmonary arteries are opened from the superior vena cava to the left upper lobe branch (Fig. 2). An incision is made in the base of the right atrial appendage but is stopped before its junction with the superior vena cava. This incision has been modified to avoid injuring the artery to the sinoatrial node. The pulmonary artery is sutured to the outside wall of the superior vena cava until the right atrial incision is

reached, transferring the suture line to the atriotomy itself. The pulmonary arteries are then enlarged with the allograft patch. A separate patch is placed within the right atrium, at the level of the limbus of the septum secundum, which will isolate superior vena caval return into the pulmonary arteries and provide an unobstructed pathway for the connection of inferior vena caval return at the time of the Fontan procedure. Prior to the insertion of this patch, the atrial septal defect is inspected and enlarged, if necessary. This is best accomplished by cutting back the coronary sinus into the left atrium. If indicated, tricuspid valve repair is also performed at this time.

The third stage: The Fontan procedure. The majority of patients following the second stage reconstruction have maintained satisfactory palliation for a considerable period of time.[5] Some, however, will experience a progressive decline in systemic arterial oxygen saturation with the development of veno-venous collaterals to the inferior vena caval circulation. Additionally, increasing systemic hypoxemia will be noted in some patients as a result of increasing growth and activity leading to an increased return of desaturated lower extremity blood with exercise. Those patients with azygous continuation of an interrupted inferior vena cava have experienced a progressive fall in oxygen saturation as venous collaterals form to the hepatic venous circulation. The development of arteriovenous malformations may constitute another important etiology for progressive hypoxemia in this group.[17] Additionally, chronic hypoxemia will stimulate growth and development of systemic arterial collaterals into the pulmonary circulation resulting in increased volume overload which may complicate the postoperative course after the Fontan procedure. For these reasons, the Fontan procedure has generally been performed at approximately 18 months to 2 years of age. Preoperative evaluation, including Doppler/echocardiography and cardiac catheterization, is essential to assess ventricular function, atrioventricular valve regurgitation, pulmonary artery size, pressure and resistance, and the presence of systemic to pulmonary artery collateral vessels. Major collaterals should be occluded by coil embolization preoperatively. The size of the left pulmonary artery is of potential

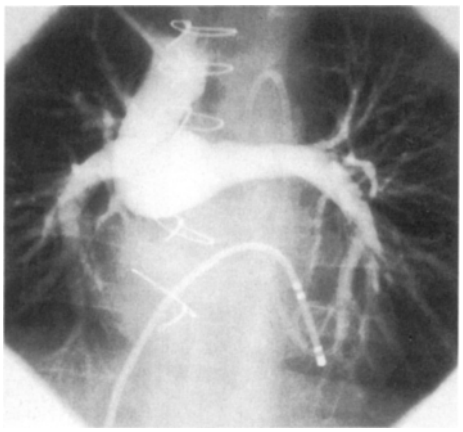

Fig. 3. Angiogram performed with contrast injected into the superior vena cava following the hemi-Fontan procedure. Unobstructed pulmonary arteries are visualized and no contrast enters the right atrium.
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concern following staged reconstruction for HLHS (Fig. 3). A dilated neoaorta following the Norwood procedure may cause compression and subsequent hypoplasia of the left pulmonary artery. In addition, the low pressure and reduced flow situation imposed by the hemi-Fontan procedure may result in a left pulmonary artery of insufficient size for the Fontan. Insertion of an endoluminal stent at the time of operation has proved to be a useful adjunct in a few of our patients with otherwise refractory hypoplasia of the left pulmonary artery. We prefer to utilize stents sparingly, and, when necessary, to place them at the final procedure when a larger size stent can be safely inserted.

Surgical technique. The heart is again exposed through a midline sternotomy, dissecting adhesions sparingly to avoid injury to the phrenic nerves. The majority of the Fontan procedures were initially performed on continuous cardiopulmonary bypass with bicaval venous return. More recently, however, single venous return is accomplished through the right atrium and the procedure is done during a brief period of circulatory arrest. The substantially reduced operative dissection required for this approach has been possible because of the preliminary connections made at the hemi-Fontan operation.[18] The entire operative procedure and, in par-

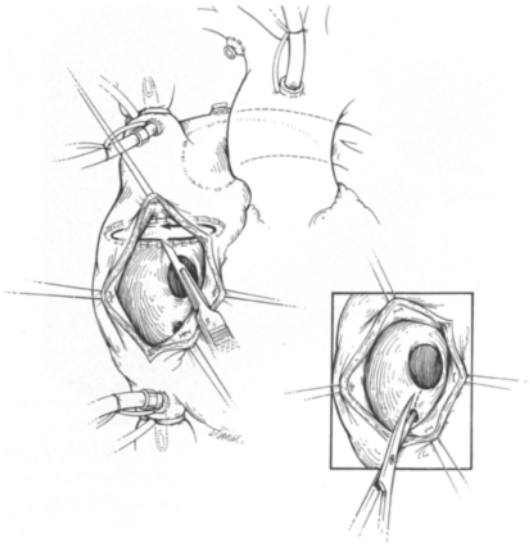

Fig. 4. The Fontan procedure is performed by first removing the previously inserted patch beneath the superior vena caval-pulmonary artery connection. The coronary sinus is cut back to enlarge the atrial septal defect if necessary (insert).
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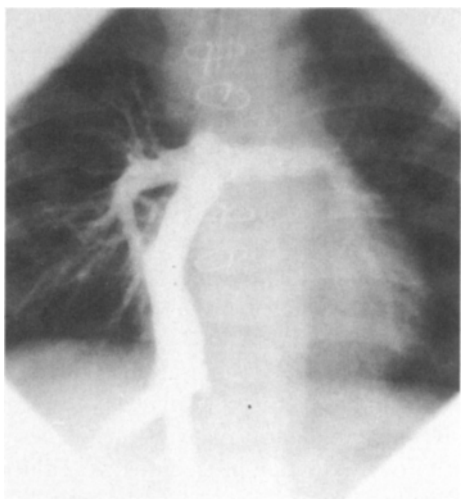

Fig. 5. Angiogram performed after the Fontan procedure demonstrating the completed cavopulmonary connection. A streamlined pathway from the inferior vena cava is shown.
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ticular, the cardiopulmonary bypass and aortic cross clamp times, have been reduced considerably (Table I). The right atrium is opened and the previously placed patch between the superior vena cava-pulmonary artery connection and the right atrium is removed (Fig. 4). A goretex patch is then fashioned to channel inferior vena caval return to the pulmonary arteries (Fig. 5). Extracardiac conduits have been used only in circumstances where a lateral tunnel cannot be constructed (e.g., anomalous pulmonary venous connection, certain heterotaxy syndromes). A fenestration of 3 to 4 mm in diameter is performed.

Table I. *Cardiopulmonary bypass and cross clamp times for the Fontan procedure*

Type of Second Stage	*CPB Time minutes*	*Cross Clamp Time minutes*
Bidirectional Glenn	111±32	59±19
Hemi-Fontan	56±10	39± 8

Results

A retrospective study was performed to evaluate the outcome of all patients with the diagnosis of HLHS who presented to C.S. Mott Children's Hospital, University of Michigan Health System, and underwent first stage reconstructive surgery with a Norwood procedure between January 1990 and August 1995.[19,20] In order to obtain an accurate assessment of the results for those patients with a single, isolated malformation, only patients with *classic* HLHS were included for analysis. For the purposes of that review, classic HLHS was defined as a right ventricular dependent circulation in association with atresia or severe hypoplasia of the aortic valve. Therefore, patients with hypoplastic left ventricles but with an otherwise adequate aortic outflow tract (e.g., transposition of the great arteries with mitral atresia) were excluded as were patients with aortic atresia or hypoplasia but with an adequate left ventricle (e.g., double inlet left ventricle with aortic atresia). This was true even though a Norwood procedure was performed. Additionally, those patients with double outlet right ventricle, atrioventricular septal defect, and atrioventricular discordance were excluded from analysis, except in the rare instance where both left ventricular and

aortic atresia or hypoplasia existed together. Patients entered into the study were divided into "standard risk" and "high risk" groups according to certain predefined criteria established from prior analyses performed at this institution.[14,20]

Definition of risk groups. In order to define the high risk population, patients presenting with coexisting conditions known to affect outcome were considered as a separate group. Considered among the high risk group were patients undergoing the Norwood procedure beyond the first month of life and those presenting with severe obstruction to pulmonary venous return. The latter was defined as the occurrence of severe hypoxemia in conjunction with radiographic evidence of pulmonary edema and an intact or nearly intact atrial septum confirmed by Doppler/echocardiography and by direct surgical or pathologic inspection. Finally, patients with significant noncardiac congenital conditions that were judged to affect or potentially affect prognosis were also excluded from the standard risk group. Such conditions included prematurity (less than 35 week gestational age), low birth weight (<2.5 kg), chromosomal anomalies, and major noncardiac anomalies such as diaphragmatic hernia. The remaining patients were considered at standard risk, but this group was anatomically diverse and, therefore, patients with coexisting anomalies of the ventricular or atrioventricular septum, visceroatrial situs defects, or atrioventricular discordance were analyzed separately. Patients were not excluded for systemic or pulmonary venous or arterial anomalies. Because prior reports from this institution failed to identify the specific morphologic subgroup (i.e., aortic atresia and mitral atresia, aortic atresia and mitral hypoplasia, aortic hypoplasia and mitral atresia, aortic hypoplasia and mitral hypoplasia) as a risk factor[14] no further anatomic subdivision of the standard risk group was undertaken for the current analysis. Patients were not stratified by preoperative condition. Therefore, the standard risk group was composed of patients with typical HLHS, including those with minor anatomic variants, who underwent primary Norwood operation within the first month of life and who did not have severe obstruction to pulmonary venous return or significant noncardiac conditions. All other patients were analyzed in the high risk group.

Clinical data. Between January 1990 and September 1998, 303 patients with classic HLHS underwent a Norwood procedure at the University of Michigan, exclusive of those patients underegoing Norwood procedures for other single ventricle lesions with systemic outflow tract obstruction. A detailed analysis of the first 158 of these patients operated upon between January 1990 and August 1995 was previously reported.[19] Between September 1995 and September 1998, an additional 145 patients with the classic form of HLHS, both standard and high risk, have undergone the Norwood procedure at our institution. Hospital survival for this latter group has been similar to those patients in the previous study. Because long term analysis is not yet available for these more recent patients, the data reported below represent that from our prior report.

Survival. Of the 158 patients undergoing the Norwood procedure, there were 120 hospital survivors (76%, 95% confidence limit [CL95]: 69%–83%). Hospital survival was significantly better among the 127 standard risk patients, 86% (CL95: 80%–92%) when compared to those in the high risk group, 42% (CL95: 25%–59%), p=0.0001. Patients were excluded from the standard risk group because of associated noncardiac congenital conditions in 17, severe pulmonary venous obstruction in 8, and age beyond 1 month at initial surgical intervention in 6. Further analysis of the high risk group revealed that hospital survival following the Norwood procedure was significantly worse among those patients with significant noncardiac congenital conditions (p=0.008) and in those with severe obstruction to pulmonary venous return (p=0.03).

There were 124 hemi-Fontan procedures performed with 121 hospital survivors, 98% (CL95: 96%–100%). There was one late death. Three of the survivors are not considered to be candidates for the Fontan procedure because of neurologic dysfunction (pre Norwood), pulmonary artery distortion unresponsive to surgical repair and endoluminal stenting, and right ventricular dysfunction following an influenza-like illness in 1 each. The latter patient has undergone suceessful cardiac transplantation.

To date, 117 patients in this group have completed the Fontan procedure.[21] Overall survival was 91% (CL95: 86%–96%). Survival was significantly better among the 65 patients who underwent a preliminary hemi-Fontan operation, 98% (CL95: 95%–100%) when compared to those undergoing a bidirectional Glenn procedure, 81% (CL95: 70%–92%), $p<.05$.

The largest decrease in survival occurred in the first month of life. Losses among survivors of the Norwood procedure prior to or related to the hemi-Fontan procedure affected primarily the high risk group and deaths occurring during the second and third years of life were primarily due to hospital death following the Fontan operation. The latter was due primarily to unrecognized stenosis or hypoplasia of the left pulmonary artery and occurred early in our experience before routine use of the hemi-Fontan procedure. Survival at 1 month, 1 year, and 5 years for the 127 patients considered at standard risk and with typical anatomy was 85 ± 4%, 80 ± 5%, and 69 ± 8%, while survival for those with anatomic variants was 86 ± 11%, 71 ± 17%, and 71 ± 17%, respectively. Survival for high risk patients was 61 ± 14%, 20 ± 40%, and 20 ± 40% at 1 month, 1 year, and 2 years, respectively.

Morbidity. Significant or potentially significant morbid conditions were noted in 25 of the 120 hospital survivors, 21% (CL95: 14%–28%). Morbidity related to the cardiovascular system was noted in 12 patients. Morbid conditions included: dysrhythmias in 5 patients (3 with permanent pacemakers, 1 with supraventricular tachycardia requiring amiodarone and 1 with sick sinus syndrome), left pulmonary artery occlusdion in 4 patients (patency restored in 3), chronic pleural effusions in 2, pulmonary artery hypertension requiring oxygen therapy in 1 patient in whom both the Norwood and hemi-Fontan procedures were performed after awaiting a transplant for 7 months at another institution, and dilated cardiomyopathy requiring transplantation at age 27 months in 1 patient. One patient had both dysrhythmias and pulmonary artery occlusion. Mechanical ventilation was required in 4 patients, with only one late survivor. Hemidiaphragm paralysis was noted in 7 patients, 6 of whom underwent plication.

Neurodevelopmental outcome. Late neurodevelopmental outcome was assessed in 49 patients following the Fontan procedure in a recent study from our institution.[22] Wechsler Preschool and Primary Scales of Intelligence-Revised were used for patients between 34 and 72 months of age and Wechsler Intelligence Scales for Children for those between 73 and 96 months. Mean verbal, performance, and overall test scores were normal for the 25 patients in this study with HLHS, although slightly lower than those patients with other single ventricle lesions. For both groups, verbal and communication skills were significantly and consistently better than motor and performance skill.

Discussion

The term hypoplastic left heart syndrome, first used by Hauck and Nadas in 1958, has remained a clinically useful description of a constellation of cardiac malformations all characterized by underdevelopment or absence of the left ventricle. The ability to make a precise diagnosis in the newborn and the introduction of prostaglandin E_1 to maintain patency of the ductus arteriosus to support the systemic circulation allowed the development of palliative surgical approaches. The majority of babies with HLHS are otherwise normal newborns. Only 5% are premature or small for gestational age. Noncardiac malformations coexist in 12% and are major in only 2–3%. Without treatment, 95% are dead within the first month of life. The postnatal physiology is characterized by an obligatory left to right shunt at atrial level and the need for the right ventricle to supply both the parallel systemic and pulmonary circulations.[22] The coronary and brachiocephalic vessels are perfused in a retrograde fashion. Alteration in hemodynamic status is determined by 3 major factors: (1) the expected fall in pulmonary vascular resistance, (2) constriction of the ductus arteriosus, (3) and the adequacy of the atrial communication. Stabilization of the newborn with HLHS is, consequently, dependent on the status and/or ability to favorably influence these factors.

There is no doubt that staged reconstruction now offers survival for patients with HLHS that equals that currently reported for many other forms of complex congenital cardiac malformations. In our

series, the actuarial survival rate, including hospital mortality for each operative procedure, for those patients considered at standard risk is approximately 70% at 5 years. These data compare favorably to those for patients with other forms of single ventricle, pulmonary atresia with intact ventricular septum, and even for some complex forms of tetralogy of Fallot. High risk patients, on the other hand, did not fare as well. For these patients, it may be reasonable to conclude that cardiac transplantation should be offered instead of staged reconstruction. However, the often grave and unstable condition of most of these patients would clearly preclude them from transplantation as a viable option. In an effort to improve the results among this group of patients, those presenting with severe obstruction to pulmonary venous return now undergo preliminary atrial septectomy, either percutaneous or operative, and the Norwood procedure is postponed until evidence of diminished pulmonary vascular resistance and lung fields free of edema are seen. Because the greatest risk for death occurs during the first month of life, efforts to reduce perioperative Norwood mortality would be expected to result in improved late survival. Survival following the hemi-Fontan procedure has been excellent and late sudden death unrelated to subsequent operations has been rare. Additionally, mortality associated with the Fontan procedure occurred almost entirely with the earliest group of patients coming to the third stage. This risk has been substantially reduced in the more recent patients and survival was 98% when the second stage procedure was a hemi-Fontan, most probably secondary to more complete relief of branch pulmonary artery stenosis in addition to shorter bypass and cross clamp times.

It is often difficult to be certain that postoperative neurologic and/or cardiovascular conditions are a result of any given operation or intervention because many of these patients are gravely ill upon presentation, prior to any procedures.[24–26] Nonetheless, previous attempts to correlate preoperative condition to outcome failed to reveal an association and many patients who were moribund prior to their Norwood procedure achieved excellent late results.[14] The effect of the operative procedures, including the use of deep hypothermia and circulatory arrest, are unclear but are currently undergoing detailed evaluation. In a recent report from our institution, Goldberg and colleagues found that neurodevelopmental outcome was normal in the majority of patients evaluated after the Fontan procedure.[22] Although the group of patients with HLHS fared slightly less well than other patients with single ventricle lesions, most remained within the normal range. Morbidity related to the cardiovascular and respiratory systems will undoubtedly always be a part of these complex surgical interventions, but the modifications noted earlier have reduced their impact.

Conclusions

Staged reconstruction for neonates born with HLHS offers a viable form of treatment with a good intermediate-term outcome. Survival has substantially improved and morbidity surrounding various interventions has been significantly reduced. Although it appears that most survivors grow and develop normally, it is essential to continually evaluate these patients to assess late quality of life. Current efforts aimed at reducing mortality following the Norwood operation by providing a more stable circulation and at eliminating the risk factors for the Fontan procedure would be expected to result in continued improvements in late survival.

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