ANNALS OF SURGERY
Vol. 224, No. 3, 387–395

Staged Reconstruction for Hypoplastic Left Heart Syndrome

Contemporary Results

Edward L. Bove, M.D.* and Thomas R. Lloyd, M.D.†

From the Section of Thoracic Surgery, Department of Surgery, and the Division of Pediatric Cardiology, Department of Pediatrics,† C. S. Mott Children's Hospital, University of Michigan School of Medicine, Ann Arbor, Michigan*

Objective

The authors review their experience with staged reconstructive surgery for hypoplastic left heart syndrome (HLHS) and assess current outcome for this condition.

Summary Background Data

Once considered a uniformly fatal condition, the outlook for newborns with HLHS has been altered dramatically with staged reconstructive procedures. Refinements in operative technique and perioperative management have been largely responsible for this improved outlook.

Methods

The authors reviewed their experience with 158 consecutive patients undergoing stage 1 reconstruction with a Norwood procedure from January 1990 to August 1995. All patients had classic HLHS, defined as a right ventricular dependent circulation in association with atresia or severe hypoplasia of the aortic valve.

Results

There were 120 hospital survivors. Among the 127 patients considered at standard risk, survival was significantly higher than that for the 31 patients with important risk factors. Adverse survival was associated most strongly with significant associated noncardiac congenital conditions and severe preoperative obstruction to pulmonary venous return. Second-stage reconstruction with the hemi-Fontan procedure was performed in 106 patients, with 103 hospital survivors and one late death. Three of the late survivors were not considered candidates for the Fontan procedure. To date, the Fontan procedure has been completed in 62 patients, with 53 survivors. Deaths after the Fontan procedure occurred early in our experience and were mostly secondary to left pulmonary artery stenosis or hypoplasia. Significant or potentially significant morbid conditions were noted in 25 of the 120 hospital survivors. Neurologic conditions were found in 6% and cardiovascular conditions in 10%, including dysrhythmia, left pulmonary artery thrombosis, and chronic pleural effusions. Among the patients considered at standard risk with typical anatomy, actuarial survival was 69 ± 8% at 5 years. Survival was 71 ± 17% at 5 years for standard risk patients with variant anatomy and 58 ± 9% for the entire cohort of 158 patients. The largest decrease in survival occurred in the first month of life and late deaths primarily affected those patients in the high-risk group.

Conclusions

Staged reconstruction has significantly improved the intermediate-term outlook for patients with HLHS. Factors addressing improvements in early (<1 month) first-stage survival would be

expected to add significantly to an overall improved late outcome. Currently employed refinements in operative technique are associated with eliminating or reducing pulmonary artery distortion and dysrhythmia.

Few congenital heart malformations have raised as many surgical, ethical, social, and economic issues as the therapy for infants born with hypoplastic left heart syndrome (HLHS). Most often, the newborn with this condition is otherwise healthy, and the prevalence of serious associated noncardiac conditions, although still not rare, is relatively low. Untreated, the prognosis for the newborn with HLHS is one of certain death, usually within hours or days of birth.[1] However, the outlook for this otherwise uniformly fatal condition has been altered dramatically in the past decade with the successful development of staged reconstructive procedures.[2] Refinements in operative technique, diagnostic imaging, and perioperative management have led the way to this significant improvement in outcome.

Although the prognosis for the newborn with HLHS is no longer one of certain death, many issues surrounding the surgical treatment remain poorly defined. Although the diagnosis of HLHS is useful to categorize the basic pathophysiology of the malformation, a great many variations exist in patient, procedural, and morphologic conditions among these patients, which may have an important influence on outcome. Additionally, improvements have evolved rapidly and current results may not reflect those reported only a few years ago. Many prior reports have included patients with cardiac anomalies that, although physiologically similar to HLHS, represent conditions that are substantially different and consequently may significantly influence the overall outcomes. Therefore, this retrospective review was conducted to determine the prognosis of contemporary patients with classic HLHS presenting to a single center.

METHODS

Study Design

The study used a retrospective cohort design to evaluate the outcome of all patients with the diagnosis of HLHS who presented to C. S. Mott Children's Hospital, University of Michigan Medical Center, and underwent first stage reconstructive surgery with a Norwood procedure. 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 this review, classic HLHS was defined as a right ventricular-dependant 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 if 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 in which 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.[3,4]

Definition of Risk Groups

To define the high-risk population, patients with coexisting conditions known to affect outcome were considered 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 also were excluded from the standard risk group. Such conditions included prematurity (<35-week gestational age), low birth weight (<2.5 kg), and chromosomal anomalies. 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

Presented at the 116th Annual Meeting of the American Surgical Association, April 18–20, 1996, Phoenix, Arizona.

Address reprint requests to Edward L. Bove, M.D., 1500 East Medical Center Drive, F7830 Mott Hospital, Box 0223, Ann Arbor, MI 48109.

Accepted for publication April 22, 1996.

atresia and mitral hypoplasia, aortic hypoplasia and mitral atresia, aortic hypoplasia and mitral hypoplasia) as a risk factor, no further anatomic subdivision of the standard risk group was undertaken for the current analysis.[3,4] 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

To identify all patients with HLHS seen at C. S. Mott Children's Hospital, University of Michigan Medical Center, between January 1990 and August 1995, the Pediatric Cardiology Diagnostic File and the Pediatric Cardiovascular Surgery Database were searched for the diagnosis of hypoplastic left ventricle and Norwood procedure, respectively. Records were reviewed for all identified patients whose birth dates fell between January 1, 1990 and August 31, 1995. Patients whose initial operation was performed at another institution were eliminated from the study. The age at most recent follow-up examination or at death, the dates of the Norwood, hemi-Fontan, and Fontan procedures, and any additional operations or interventions were recorded. Conditions causing (or potentially causing) postoperative morbidity were noted, with particular emphasis on those conditions resulting in neurologic or cardiopulmonary compromise. Delayed sternal closure and early reoperation for bleeding were not analyzed as separate risk factors.

Statistical Analysis

Survival analysis was performed by the product-limit (Kaplan-Meier) method, with survival calculated at 30-day intervals for the first year of life and at 180-day intervals thereafter. Hospital deaths were considered as any deaths during the hospital stay, regardless of duration. Survival comparisons were performed by contingency analysis and $p < 0.05$ was considered to represent statistical significance. The continuity correction was employed for comparisons with one degree of freedom. Survival estimates calculated as proportions are presented with their 70% confidence limits. Those calculated by the actuarial method are presented with standard errors.

Table 1. PATIENT GROUPS*

Group	No.
Standard risk	127
High risk	31
Noncardiac congenital condition	17
Prematurity/low birth weight	4
Chromosomal anomaly	5
Galactosemia	1
Absent corpus callosum	1
Anal atresia	1
Chronic renal failure	1
Multiple limb defects	1
Congenital diaphragmatic hernia	1
Recurrent left atrial tumor	1
Hypothyroidism and portal fibrosis	1
Pulmonary venous obstruction	8
Age > 1 mo at Norwood	6

* n = 158.

RESULTS

Patient Groups

A total of 164 patients with the diagnosis of HLHS were identified. No cardiac operation was performed in 5 patients, and 1 did not receive a Norwood procedure, leaving 158 patients with classic HLHS for analysis (Table 1). Among the 158 patients, 127 were considered at standard risk. Within this group, there were 14 patients with anatomic variants. In each case, the basic pathologic anatomy, as defined previously for classic HLHS (hypoplastic left ventricle with dominant right ventricle and aortic atresia or hypoplasia), was present. However, the variation from the typical anatomy (Table 2) prompted separate analysis. The remaining 31 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.

Additional Operations/Interventions

In addition to the 158 primary Norwood operations, hemi-Fontan procedures have been performed in 106 patients and Fontan procedures in 62 patients. Additional cardiovascular interventions (Table 3) performed either in the catheterization laboratory or in conjunction with subsequent staged procedures included coil embolization of systemic aortopulmonary collateral arteries (16), pulmonary artery balloon dilatation or endoluminal stenting (12), and balloon dilatation of residual coarctation (6). Tricuspid valvuloplasty was performed at the time of hemi-Fontan or Fontan procedure in eight

Table 2. ADDITIONAL OPERATIONS/ INTERVENTIONS

Operation/Intervention	No.
Cardiovascular	58
Coil embolization AP collaterals	16
PA balloon/stent insertion	12
Balloon dilate residual coarctation	6
Tricuspid valvuloplasty	8
Atrial septectomy	4
Shunt revision after Norwood	3
PA augmentation/thrombectomy	3
Pacemaker insertion	3
Central shunt after hemi-Fontan	1
Fontan takedown	1
Heart transplant	1
Noncardiovascular	20
Hemidiaphragm plication	6
Tracheostomy	4
Drainage pericardial effusion	6
Pleurodesis	3
Lobectomy	1

AP = aortopulmonary; PA = pulmonary artery.

patients, an additional atrial septectomy was required in four (2 prior to the Norwood), shunt revision after the Norwood procedure in three, left pulmonary artery augmentation/thrombectomy in three, permanent pacemaker insertion in three, and central shunt following hemi-Fontan procedure, Fontan takedown, and heart transplantation in one patient each. Noncardiovascular procedures included hemidiaphragm plication (6), tracheostomy (4), drainage of pericardial effusion (6), pleurodesis (3), and lobectomy for necrotizing pneumonia (1).

Circulatory assistance (extracorporeal membrane oxygenation) was required in eight patients. Two patients were supported before the Norwood procedure, with one survivor; two were supported after the Norwood procedure, with one survivor; two were supported after the hemi-Fontan procedure, with one survivor; and two were supported after the Fontan procedure, with no survivors.

Survival

Of the 158 patients undergoing the Norwood procedure, there were 120 hospital survivors (76%, 70% confidence limit [CL_{70}]: 72%–79%). Hospital survival was significantly better among the 127 standard risk patients (86%, CL_{70}: 82%–89%) when compared with those in the high-risk group (42%, CL_{70}: 33%–51%; $p = 0.0001$). Among the standard risk group, there was no difference in hospital survival for those with typical anatomy (87%, CL_{70}: 83%–90%) and those with anatomic variants (79%, CL_{70}: 65% to 88%; p = not significant). Further analysis of the high-risk group revealed that hospital survival after 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 106 hemi-Fontan procedures performed, with 103 hospital survivors (97%; CL_{70}: 95%–98%). 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 after an influenza-like illness in one each. The latter patient has undergone successful cardiac transplantation. All remaining 99 patients have either undergone or are considered candidates for the Fontan procedure. To date, the Fontan procedure has been completed in 62 patients, with 53 hospital survivors (86%; CL_{70}: 80%–90%). One hospital survivor died 15 months after the Fontan procedure secondary to pneumonia.

Actuarial survival estimates are shown in Figures 1 and 2. The largest decrease in survival occurred in the first month of life. Losses in survivors of the Norwood procedure before or related to the hemi-Fontan procedure affected primarily the high-risk group, and deaths occurring during the second and third years of life primarily were due to hospital death after the Fontan operation. The latter was due primarily to unrecognized stenosis or hypoplasia of the left pulmonary artery and occurred early in our experience. Among the entire patient

Table 3. POSTOPERATIVE MORBIDITY

Condition	Number
Neurologic	7
Thromboembolic stroke	2
Absent corpus callosum	1
Developmental delay	3
Feeding problems/FTT	1
Cardiovascular	12*
Dysrhythmias	5
Left PA occlusion	4
Chronic pleural effusions	2
PA hypertension	1
Cardiomyopathy	1
Respiratory	11
Mechanical ventilation	4
Hemidiaphragm paralysis	7

FTT = failure to thrive; PA = pulmonary artery.
* One patient had two conditions.

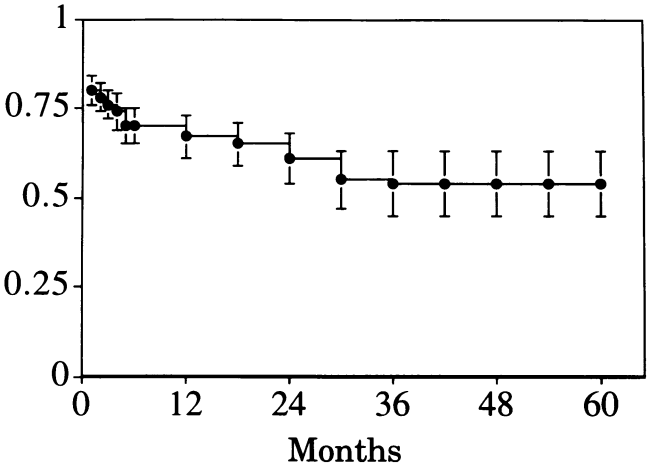

Figure 1. Actuarial survival for the entire study group of 158 patients.

Table 4. COEXISTING ANATOMIC VARIANTS IN STANDARD RISK PATIENTS*

Variant	Number
AV septal defect	6
AV septal defect, DORV	2
AV septal defect, CHB	1
DORV	2
DORV, dextrocardia, situs inversus	1
TGA (SLD), dextrocardia	1
Apical VSD	1

AV = atrioventricular; DORV = double outlet right ventricle; CHB = complete heart block; TGA = transposition of the great arteries; VSD = ventricular septal defect.
* n = 14. All patients also had hypoplastic left ventricle with dominant right ventricle and aortic atresia or hypoplasia.

group, actuarial survival rates were 80 ± 4%, 67 ± 6%, and 58 ± 9% at 1 month, 1 year, and 5 years, respectively. Survival rates at the same time intervals for the 127 patients considered at standard risk and with typical anatomy were 85 ± 4%, 80 ± 5%, and 69 ± 8%, whereas survival rates for those with anatomic variants were 86 ± 11%, 71 ± 17%, and 71 ± 17%. Survival rates for high-risk patients were 61 ± 14%, 20 ± 40%, and 20 ± 40% at 1 month, 1 year, and 2 years, respectively.

Morbidity

Significant or potentially significant morbid conditions (Table 4) were noted in 25 of the 120 hospital survivors (21%; CL_{70}: 17%–25%). Neurologic conditions were noted in seven patients (6%; CL_{70}: 4%–9%), including two patients with thromboembolic strokes, one patient with absent corpus callosum and severe developmental delay, three patients with developmental delay, and one patient with feeding problems and failure to thrive in whom neurologic involvement is suspected. Morbidity related to the cardiovascular system was noted in 12 patients (10%; CL_{70}: 8%–13%). Morbid conditions included dysrhythmia in five patients (3 with permanent pacemakers, 1 with supraventricular tachycardia requiring amiodarone, and 1 with sick sinus syndrome), left pulmonary artery occlusion in four patients (patency restored in 3), chronic pleural effusions in two patients, pulmonary artery hypertension requiring oxygen therapy in one 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 one patient. One patient had both dysrhythmia and pulmonary artery occlusion. Mechanical ventilation was required in four patients, with only one late survivor. Hemidiaphragm paralysis was noted in seven patients, six of whom underwent plication.

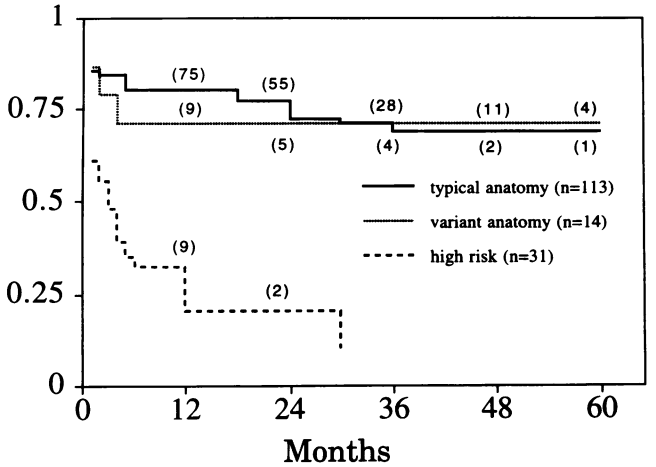

Figure 2. Actuarial survival analyzed for standard and high-risk groups separately. The standard risk group is subdivided further into those with typical anatomy and those with variant anatomy (see text for details).

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. In 1980, Norwood and associates[2] first reported the successful application of a palliative operation that provided unob-

structed systemic blood flow from the right ventricle to the aorta through a conduit and restricted pulmonary flow through a systemic to pulmonary artery shunt. Subsequent surgical procedures were designed to facilitate the transition to a physiologically normal circulation with the connection of the superior vena cava (second stage) and the inferior vena cava (third stage) to the undivided pulmonary arteries. Although the early results were discouraging, many modifications of each of the procedures have been developed and have resulted in a dramatic reduction in mortality.[3-5] Concomitant improvements in the knowledge of perioperative physiology also have been instrumental in the care of these patients, in whom even minor changes in the delicate balance between pulmonary and systemic vascular resistance may be associated with the rapid development of an unstable circulation.[6]

Norwood Procedure

Although the basic principles for each of the three stages remain consistent, modifications in operative techniques continue to result in improved outcomes.[7-9] The essential elements of the Norwood operation are the provision of unobstructed systemic and coronary blood flow with aortic arch augmentation, relief of pulmonary venous obstruction by atrial septectomy, and control of pulmonary blood flow through a restrictive systemic to pulmonary artery shunt. The reconstruction of the aorta and its association with the pulmonary valve must be performed without excessive dilatation of the allograft patch to avoid compression of the left pulmonary artery.[10] Early in our experience, this problem resulted in some—often subtle—left pulmonary artery obstruction that only became hemodynamically significant after the Fontan procedure, when the entire cardiac output passes through the lungs. Obstruction to pulmonary artery blood flow was believed to be the most significant contributing factor to death among the early patients in this series undergoing the Fontan procedure. More streamlined tailoring of the allograft patch at the time of the Norwood procedure and routine augmentation of the left pulmonary artery at the time of the hemi-Fontan procedure has improved substantially this important problem in the more recent patients.[7]

Reconstruction of the aorta to provide unobstructed coronary artery flow presents a technical challenge when the ascending aorta is less than 2 to 3 mm in diameter. However, careful alignment of the aorta with the proximal pulmonary artery to avoid rotation or kinking is possible, even in patients with an extremely diminutive ascending aorta. Prior analyses from this and other institutions failed to identify the size of the ascending aorta as a risk factor for death.[3,11]

The control of pulmonary blood flow remains the most significant issue surrounding the Norwood operation and is the factor associated most often with early death. The dynamic balance between the systemic and pulmonary vascular resistance is altered easily and depends on many factors. Although the size of the shunt itself is one major element, this must be viewed in the context of the overall patient condition, particularly the status of the pulmonary vascular resistance. Older age at operation is likely to be associated with elevated resistance, and postoperative pulmonary blood flow may be inadequate if too small a shunt is used. A similar, but usually transient, situation exists in patients with pulmonary edema resulting from excessive pulmonary blood flow or obstruction to pulmonary venous return. Conversely, a large shunt will provide too much flow when resistance is low, and the resultant pulmonary overcirculation will lead to low cardiac output, systemic acidosis, and early death. Because postoperative condition is considerably more stable when right ventricular volume overload is avoided by using a small shunt, the optimal patient undergoing the Norwood procedure should have low pulmonary vascular resistance and no increase in extravascular lung water. Thus, the patients undergoing first-stage reconstruction beyond the neonatal period and those with obstructed pulmonary venous return have posed a significantly greater risk. The group of patients presenting with prematurity and low birth weight also have posed an increased risk predominantly from the inability to sufficiently limit pulmonary flow, even when 3-mm polytetrafluoroethylene shunts are used. Recently, we have used standard Blalock-Taussig shunts in patients weighing less than 2.5 kg and have noted more appropriate pulmonary blood flow, possibly because the smaller subclavian artery acts as the flow regulating vessel. This modification recently has been used successfully in five consecutive patients weighing less than 2.1 kg.

Hemi-Fontan Procedure

Removal of the ventricular volume overload resulting from the systemic shunt has proven to be an important step in reducing mortality from the Fontan procedure.[12-14] However, when small shunts are used at the first stage, pulmonary blood flow rapidly will become insufficient with patient growth and the second stage will be required at an early age. This has not resulted in an increased risk, however, and hemi-Fontan procedures have been performed successfully as early as 1 month of age.[12] The salutary effect of the hemi-Fontan procedure probably results from the fact that after this operation, patients generally remain well palliated, free of risk factors, until they reach an age when they are more suitable

candidates for the Fontan procedure. Therefore, it is essential that all risk factors be addressed at the second stage, including tricuspid regurgitation, pulmonary artery hypoplasia, residual obstruction to pulmonary venous return, and systemic outflow tract obstruction. In addition, a potential connection for the inferior vena caval return must be provided at the subsequent Fontan procedure to simplify that stage and avoid the need for additional dissection, which may damage the sinus node or its arterial supply.

Fontan Procedure

After a properly constructed hemi-Fontan operation, the final stage of the reconstruction should be the simplest procedure. It remains our preference to construct an intra-atrial lateral tunnel, channeling the inferior vena cava to the pulmonary arteries through the connection made at the previous operation.[7,15,16] Removal of the temporary wall separating the superior vena cava to pulmonary artery anastomosis from the right atrium will result in a large unobstructed pathway. Therefore, the operative dissection is kept to a minimum, which reduces blood loss and potential injury to the conduction system and phrenic nerves. Total cardiopulmonary bypass time and aortic cross clamp time also are minimized, resulting in less perioperative edema and pulmonary dysfunction. Since adopting these strategies at our institution, the risk of the Fontan procedure has decreased dramatically and the postoperative courses have been mostly uncomplicated.

Postoperative Morbidity

Any analysis of the results of a surgical strategy of staged reconstruction for neonates with HLHS must include an assessment of morbidity. It often is difficult to be certain that postoperative morbid conditions are a result of any given operation or intervention because many of these patients were gravely ill on presentation, before any procedures.[17–19] Nonetheless, previous attempts to correlate preoperative condition to outcome failed to reveal an association, and many patients who were moribund before their Norwood procedure achieved excellent late results.[3] Although postoperative neurologic conditions were noted in 6% of hospital survivors, some were congenital and others likely were a result of preoperative shock. The effect of the operative procedures, including the use of deep hypothermia and circulatory arrest, are unclear but currently are undergoing detailed evaluation at our institution. Morbidity related to the cardiovascular and respiratory systems undoubtedly will always be a part of these complex surgical interventions, but the aforementioned modifications have significantly reduced their impact.

Survival

There is no doubt that staged reconstruction offers survival for patients with HLHS that equals that currently reported for many other forms of complex congenital cardiac malformations. In this series, the actuarial survival rate for those patients considered at standard risk, including hospital mortality for each operative procedure, 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. However, high-risk patients 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 clearly would 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 currently undergo preliminary atrial septectomy alone, and the Norwood procedure is postponed until evidence of diminished pulmonary vascular resistance and lung fields free of edema are seen. Patients weighing less than 2.5 kg receive a standard Blalock-Taussig shunt, which appears to limit pulmonary blood flow to a degree better tolerated in very small birth weight babies. 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 after 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 entirely with the earliest group of patients coming to the third stage. This risk has been reduced substantially in the more recent patients.

CONCLUSIONS

Staged reconstruction for neonates born with HLHS offers a viable form of treatment with a good intermediate-term outcome. Survival has improved substantially and morbidity surrounding various interventions has been reduced significantly. 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 after the Norwood operation by providing a more stable circulation and at eliminating the risk factors for the Fon-

tan procedure would be expected to result in continued improvements in late survival.

References

1. Gutgesell HP, Massaro TA. Management of hypoplastic left heart syndrome in a consortium of university hospitals. Am J Cardiol 1995; 76:809–811.
2. Norwood WI, Kirklin JK, Sanders SP. Hypoplastic left heart syndrome: experience with palliative surgery. Am J Cardiol 1980; 45: 87–91.
3. Iannettoni MD, Bove EL, Mosca RS, et al. Improving results with first-stage palliation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 1994; 107:934–940.
4. Lloyd TR. Prognosis of the hypoplastic left heart syndrome. Progr Pediatr Cardiol 1996; 5:57–64.
5. Meliones JN, Snider AR, Bove EL, et al. Longitudinal results after first-stage palliation for hypoplastic left heart syndrome. Circulation 1990; 82(suppl IV):IV-151–IV-156.
6. Barnea O, Austin EH, Richman B, Santamore WP. Balancing the circulation: theoretic optimization of pulmonary/systemic flow ratio in hypoplastic left heart syndrome. J Am Coll Cardiol 1994; 24: 1376–1381.
7. Bove EL, Mosca RS. Surgical repair of the hypoplastic left heart syndrome. Progr Pediatr Cardiol 1996; 5:23–35.
8. Jonas RA, Lang P, Hansen D, et al. First-stage palliation of hypoplastic left heart syndrome: the importance of coarction and shunt size. J Thorac Cardiovasc Surg 1986; 92:6–13.
9. Pigott JD, Murphy JD, Barber G, Norwood WI. Palliative reconstructive surgery for hypoplastic left heart syndrome. Ann Thorac Surg 1988; 45:122–128.
10. Alboliras ET, Chin AJ, Barber G. Pulmonary artery configuration after palliative operations for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 1989; 97:878–885.
11. Helton JG, Aglira BA, Chin AJ, et al. Analysis of potential anatomic or physiologic determinants of outcome of palliative surgery for hypoplastic left heart syndrome. Circulation 1986; 74(suppl I): I-70–I-76.
12. Bradley SM, Mosca RS, Hennein HA, et al. Bidirectional superior cavopulmonary connection in young infants. Circulation (in press).
13. Pridjian AK, Mendelsohn A, Lupinetti FM. Usefulness of the bidirectional Glenn procedure as staged reconstruction for the functional single ventricle. Am J Cardiol 1993; 71:959–962.
14. Murdison KA, Baffa JM, Farrell PE Jr, et al. Hypoplastic left heart syndrome: outcome after initial reconstruction and before modified Fontan procedure. Circulation 1990; 82(suppl IV):IV-199–IV-207.
15. Norwood WI, Jacobs ML, Murphy JD. Fontan procedure for hypoplastic left heart syndrome. Ann Thor Surg 1992; 54:1025–1030.
16. Farrell PE Jr, Chang AC, Murdisen KA, et al. Outcome and assessment after the modified Fontan procedure for hypoplastic left heart syndrome. Circulation 1992; 85:116–122.
17. Rogers BT, Msall ME, Buck GM, et al. Neurodevelopmental outcome of infants with hypoplastic left heart syndrome. J Pediatr 1995; 126:496–498.
18. Glauser TA, Rorke LB, Weinberg PM, Clancy RR. Congenital brain anomalies associated with the hypoplastic left heart syndrome. Pediatrics 1990; 85:984–990.
19. Glauser TA, Rorke LB, Weinberg PM, Clancy RR. Acquired neuropathologic lesions associated with the hypoplastic left heart syndrome. Pediatrics 1990; 85:991–1000.

Discussion

DR. LEONARD L. BAILEY (Loma Linda, California): I rise mainly as a friend and fan of Ed Bove, and I want to congratulate him and the team that he works with at Ann Arbor for the splendid report that you have just heard. I think few here today understand the real magnitude of effort that has gone into this report, and it is for that reason among others that it belongs on this program.

Dr. Bove has carried Norwood's effort to its highest plane yet reported, from what we can see here today. Clearly a majority of babies with hypoplastic left heart syndrome can be salvaged in the short term. The troublesome reality remains that no matter how the data are arranged, nearly 40% of the babies with hypoplastic left hearts who have been operated on at Michigan are no longer in existence, and approximately 20% of that cohort of babies have comorbid features that may preclude additional intervention. Also, only approximately a third of the babies have achieved the ultimate (or final stage) Fontan status. So the jury may still be out on the intermediate or long-term outcome after this strategy of intervention for hypoplastic left heart syndrome.

By comparison, the Loma Linda experience with transplantation suggests that 75% of babies lucky enough to get a transplant during the first 30 days of life will still be living 10 years later. So that is, perhaps, the standard against which to measure reconstructive outcomes. We clearly have insufficient donors, so the babies cannot all be addressed by transplantation; hence, the relevance of this report on staged reconstruction.

I have a couple of questions for Dr. Bove. Are the Fontan procedures incomplete, the so-called fenestrated Fontan, or are they complete Fontans? Are the babies pink or a bit cyanotic at the end of the long stretch? I notice that only one baby in this cohort converted over to transplantation. You might want to talk a little bit about how many others might yet be favorable for transplantation, if any.

In our whole experience over 10 years of transplantation, we have only transplanted one baby that had a previous Norwood. It does not appear that very many (if they fail Norwood) would ever get to transplantation. Unfortunately, the Norwood may not be a very suitable bridge to transplantation.

DR. JOHN W. HAMMON (Winston-Salem, North Carolina): I rise to congratulate Dr. Bove for the excellent results achieved in this premiere article on the first-stage reconstruction of hypoplastic left heart syndrome using the Norwood procedure. Dr. Bove has added many technical details including postoperative management in this difficult group of patients and his results truly demonstrate his outstanding efforts. I do have a couple of questions that I would like to address to Dr. Bove in follow-up from the previous discussion.

There has been evidence in some series that the size of the ascending aorta in classic hypoplastic left heart syndrome determines in some way the efficacy of the repair by providing potential obstruction to coronary blood flow. I wonder if Dr. Bove would address that question.

The second is, with the very poor long-term survival rate in the complicated group of patients with other anomalies, does