Current outcomes and risk factors for the Norwood procedure

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Objective: Tremendous strides have been made in the outcomes for hypoplastic left heart syndrome and other functional single-ventricle malformations over the past 25 years. This progress relates primarily to improvements in survival for patients undergoing the Norwood procedure. Previous reports on risk factors have been on smaller groups of patients or collected over relatively long periods of time, during which management has evolved. We analyzed our current results for the Norwood procedure with attention to risk factors for poor outcome.

Methods: A single-institution review of all patients undergoing a Norwood procedure for a single-ventricle malformation from May 1, 2001, through April 30, 2003, was performed. Patient demographics, anatomy, clinical condition, associated anomalies, operative details, and outcomes were recorded.

Results: Of the 111 patients, there were 23 (21%) hospital deaths. Univariate analysis revealed noncardiac abnormalities (genetic or significant extracardiac diagnosis, $P = .0018$), gestational age ($P = .03$), diagnosis of unbalanced atrioventricular septal defect ($P = .017$), and weight of less than 2.5 kg ($P = .0072$) to be related to hospital death. On multivariate analysis, only weight of less than 2.5 kg and noncardiac abnormalities were found to be independent risk factors. Patients with either of these characteristics had a hospital survival of 52% (12/23), whereas those at standard risk had a survival of 86% (76/88).

Conclusions: Although improvements in management might have lessened the effect of some of the traditionally reported risk factors related to variations in the cardiovascular anatomy, noncardiac abnormalities and low birth weight remain as a future challenge for the physician caring for the patient with single-ventricle physiology.

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Until only a quarter century ago, hypoplastic left heart syndrome (HLHS) was a uniformly fatal condition. Without early intervention, 95% of affected infants died within the first month of life.¹ The outlook has improved dramatically since Norwood and colleagues² reported the first successful staged palliative reconstructive operations for infants with HLHS in 1983 and Bailey and associates³ introduced cardiac transplantation for the disease in 1986. However, despite improvements in survival, HLHS continues to be the most common anomaly resulting in death within the first year of life in the United States.⁴ Most of the mortality associated with staged repair occurs during the first stage, the Norwood

Abbreviations and Acronyms

CPB	= cardiopulmonary bypass
DHCA	= deep hypothermic circulatory arrest
FSV	= functional single ventricle
HLHS	= hypoplastic left heart syndrome
RCP	= regional cerebral perfusion

procedure. Therefore the elucidation of preoperative risk factors associated with higher mortality could lead to better identification of those infants more likely to benefit from transplantation rather than staged repair.

Many authors have previously reported risk factors for the Norwood procedure. Despite these numerous reports, there exists no consensus on which risk factors are significant. Forbess and coworkers⁵ found lower preoperative pH to be a significant risk factor for stage I mortality, whereas in a previous report⁶ our group found no increased risk. Initial operations after 14 or 30 days have both been reported as a risk factor,⁶⁻⁸ whereas others have found no increased risk for those infants operated on after 14 days of life.⁹ The data are similarly ambiguous for fetal diagnosis,¹⁰⁻¹³ anatomic subtype of HLHS,^{5,6,8,10,14,15} lower operative weight,^{5,6,8,10,16-18} smaller ascending aortic diameter,^{5,6,16,17,19-21} longer circulatory arrest^{6,11,17,20} or cardiopulmonary bypass (CPB) time,^{11,17,21} noncardiac congenital anomalies,^{7,16,22,23} and the presence of moderate-to-severe tricuspid regurgitation before the operation.^{5,17,19,24} In addition to these risk factors, 3 authors have reported a higher risk of mortality associated with obstructed pulmonary venous return.^{7,8,22,25}

Many of these previously reported risk factors were derived from small cohorts of patients or collected over relatively long periods of time, during which management patterns have evolved. These factors might account for the level of disparity among risk factors described in the literature. This report describes our recent experience with the Norwood procedure for palliation of HLHS and other functional single ventricle (FSV) malformations, with a focus on identifying relevant risk factors in the current era.

Patients and Methods**Study Design**

A single-center retrospective review of the medical records of children undergoing a Norwood procedure for the correction of HLHS or other FSV malformations at the C.S. Mott Children's Hospital of the University of Michigan Health Systems from May 1, 2001, through April 30, 2003, was performed. Approval was obtained from the institutional review board before initiation of the study.

The study hypothesis was that because of improvements in operative technique and perioperative management, many of the traditional risk factors previously reported for the Norwood procedure have been overcome.

Patient Population

Between May 1, 2001, and April 30, 2003, 111 infants underwent a Norwood procedure for HLHS or other FSV malformations. The demographic data that were collected included gestational age at birth, age at initial operation, weight at operation, sex, race, cardiac anatomy, noncardiac diagnoses, and fetal diagnosis. Surrogates for condition at presentation included history of cardiac arrest or seizures, the lowest pH, and the peak creatinine levels. Preoperative pH and creatinine levels estimated adequacy of resuscitation and immediate preoperative condition. Also noted were the need for balloon atrial septostomy and the presence of pulmonary venous obstruction, as defined by significant hypoxemia in conjunction with radiographic evidence of pulmonary edema and an intact or nearly intact atrial septum confirmed by means of Doppler echocardiography and direct surgical or pathologic inspection. Echocardiographic parameters, including cardiac valve sizes and functions, aortic size and the presence of antegrade flow, and ventricular function, were all collected. Operative parameters included time in the operating room, deep hypothermic circulatory arrest (DHCA) or regional cerebral perfusion (RCP) time, CPB time, and shunt type and size. Postoperative parameters included time to chest closure and extubation, days in the intensive care unit, and days in the hospital. Time of follow-up and continuation to hemi-Fontan or Fontan procedures were also recorded. A detailed list of the potentially significant factors that were recorded is included in Appendix 1. Hospital survival was the primary outcome.

Surgical Technique

The technique used for the first stage of reconstruction was a classic Norwood procedure, as initially described by Pigott and associates.²⁶ Important modifications, including the manner of the proximal aortic anastomosis, technique and extent of the arch reconstruction, and the use of smaller shunts, have been previously detailed in a publication from our group.²² One hundred five of the patients received a systemic-to-pulmonary artery shunt, of which 66% (69/105) were 3.5 mm, 31% (33/105) were 4.0 mm, and 3% (3/105) were 3.0 mm. Six patients (median weight, 2.2 kg; range, 1.7-3.2 kg) received a right ventricle-to-pulmonary artery shunt ranging in size from 4.0 to 5.0 mm.

Statistical Analysis

Normally distributed data are expressed as means $\pm$ standard deviation. Nonnormal data are expressed as medians and ranges. Dichotomous variables were analyzed with the Fisher exact test, and continuous variables were subjected to the Student *t* test. Wilcoxon rank sum testing was used for nonnormally distributed data.

Results**Patient Population**

Of the 111 patients, there were 71 (64.0%) male and 40 (36.0%) female patients. Mean age at the time of the operation was 9 ± 5 days. Twelve (10.8%) patients were more than 14 days old at the time of their initial operation, and 2 (1.8%) of these patients were more than 30 days old. Median weight at the time of the operation was 3.2 kg (range,

TABLE 1. Chromosomal abnormalities-syndromes

Anomaly	n
Turner	2
Indeterminate*	2
Costello	1
Jacobsen	1
VACTERL	1
Alagille	1
Ritscher-Schinzel	1
McKusick-Kauffman	1
In(3)(p11.2q25.1)	1

VACTERL, vertebral/anal/cardiothoracic/tracheoesophageal fistula/renal/limb association. *Believed by genetics consultation to be significantly syndromic without specific diagnosis.

1.3-4.5 kg). Nine (8.1%) patients weighed less than 2.5 kg at the time of their initial operations. The median gestational age at birth was 39 weeks (range, 33.0-42.5 weeks). There were 90 (81%) white, 8 (7%) African-American, 6 (5%) Hispanic, and 7 (6%) "other" or "unknown" infants. One hundred (90%) of 111 patients were given diagnoses of HLHS, as defined by a right ventricle-dependent circulation with aortic atresia or hypoplasia. Five (4%) patients had tricuspid atresia, 4 (4%) had a double-inlet left ventricle, and 2 (2%) had transposition of the great arteries with an FSV. Of these patients, 3 (3%) had heterotaxy. Overall, there were 101 (91%) patients with a functional single right ventricle, whereas 10 (9%) had a functional single left ventricle.

Of the 100 patients with HLHS, there were 36 (36%) with aortic atresia and mitral atresia, 23 (23%) with aortic stenosis and mitral stenosis, 20 (20%) with aortic atresia and mitral stenosis, 1 (1%) with aortic stenosis and mitral atresia, 10 (10%) with a common atrioventricular valve orifice, and 10 (10%) with double-outlet right ventricle. The median ascending aorta size was 3.3 mm (range, 1.40-8.60 mm). Antegrade aortic flow was seen in 49 (42%) patients.

Twelve (11%) patients were identified as dysmorphic, and 34 (31%) patients underwent genetic analysis. Eleven (10%) patients were found to have abnormal chromosomes or a named genetic syndrome (Table 1). Ten (9%) patients had significant acquired or congenital extracardiac diagnoses (Table 2).

There were 68 (61%) patients in whom a fetal diagnosis was attained. The median lowest preoperative pH was 7.42 (range, 7.25-7.53), and the median preoperative pH was 7.42 (range, 7.25-7.53). The median peak creatinine level was 0.80 mg/dL (range, 0.40-5.50 mg/dL), and the median preoperative creatinine level was 0.70 mg/dL (range, 0.40-3.90 mg/dL). No patients had a history of cardiac arrest before the initial operation, but 6 (5%) had a history of seizures. Six (5%) patients required balloon atrial septos-

TABLE 2. Extracardiac diagnoses

Diagnosis	n
Renal dysplasia-insufficiency	3
Intracranial	
Absent corpus callosum	1
Cerebrovascular accident	1
Cerebral aneurysm	1
Biliary atresia	1
Pulmonary dysplasia	1
Multiple	2

tomy before the Norwood procedure to relieve pulmonary venous obstruction.

Median time in the operating room was 205 minutes (range, 149-525 minutes). Seventy-five (68%) patients underwent DHCA, whereas RCP was used in the remaining 36 (32%) patients. Median DHCA time was 37 minutes (range, 24-68 minutes). Median RCP time was 39 minutes (range, 30-69 minutes). Median CPB time was 96 minutes (range, 59-308 minutes).

Chest closure was performed after a median of 7 days (range, 1-15 days), and patients were extubated after a median of 7 days (range, 2-46 days). The median number of days in the intensive care unit for survivors was 9 (range, 3-87 days), and the average length of hospital stay for survivors was 21 days (range, 8-148 days). Median length of follow-up was 17 months (range, 0.5-33 months), with 71 (64%) patients undergoing stage II palliation. Of these 71 patients, 30 (27%) have gone on to a Fontan procedure.

Hospital Survival and Risk Factors

The overall hospital survival was 79% (88/111 patients). The most common cause of death was low cardiac output in the immediate postoperative period in 39% (9/23) of patients, followed by unexpected arrest in patients seemingly making an unremarkable recovery in 22% (5/23). Other cardiorespiratory causes of death included high pulmonary vascular resistance in a patient with a history of obstructed pulmonary venous return (n = 1), refractory arrhythmia (n = 1), severe native aortic insufficiency (n = 1), severe neoaortic insufficiency (n = 1), and severe tricuspid regurgitation (n = 1). Noncardiac causes included sepsis (n = 1), necrotizing enterocolitis (n = 1), retroperitoneal hemorrhage caused by bladder perforation (n = 1), and intracranial hemorrhage caused by cerebral aneurysm (n = 1).

Table 3 shows the variables that could not be shown to have an effect on hospital survival. Univariable analysis revealed that noncardiac abnormalities, including genetic abnormalities or significant extracardiac malformations ($P = .0018$), gestational age at operation ($P = .03$), diagnosis of unbalanced atrioventricular septal defect ($P = .017$), and weight of less than 2.5 kg ($P = .0072$) were significantly related to hospital death (Table 4).

TABLE 3. Factors not achieving statistical significance

	Survivor	Mortality	P value
Fetal diagnosis	61% (52/85)	70% (16/23)	.42
Gestational age <35 wk	0% (0/84)	9% (2/23)	.76
Sex	60 M/28 F	11 M/12 F	.07
Age >30 d	1% (1/88)	4% (1/23)	.37
Aortic atresia	58% (58/88)	39% (9/23)	.11
Obstructed pulmonary venous return	7% (6/87)	9% (2/23)	.77
Median bypass time (min)	93.5 (59-242)	110 (65-308)	.07
Median DHCA or RCP time (min)	36 (24-68)	40.5 (30-49)	.57
RVPA conduit	3% (3/88)	17% (4/23)	NS
Median MBTS diameter (mm)	3.5 (3.0-4.0)	3.5 (3.0-4.0)	NS
Median lowest preoperative Ph	7.34 (7.11-7.46)	7.34 (7.20-7.44)	.67
Median ascending aortic size (mm)	3.25 (1.4-8.2)	3.9 (1.4-8.6)	.52
Median degree of TR (0-4+)	1+ (0-3+)	1+ (0-3+)	NS

DHCA, Deep hypothermic circulatory arrest; RCP, regional cerebral perfusion; RVPA, right ventricle to pulmonary artery; MBTS, modified Blalock-Taussig shunt; TR, tricuspid regurgitation.

The only independent risk factors for hospital survival on multivariable analysis proved to be weight of less than 2.5 kg at the time of the operation ($P = .0098$; odds ratio, 7.0) and noncardiac congenital abnormalities ($P = .032$; odds ratio, 6.0; Table 5). The presence of either weight of less than 2.5 kg or a noncardiac diagnosis conveyed a hospital survival of 52% (12/23) compared with 86% (76/88) for infants at standard risk.

Discussion

Operative survival after the Norwood procedure, the first stage in surgical palliation for HLHS and other conditions resulting in FSV physiology, has improved steadily since the early 1980s because of advancements in perioperative care and improvements in operative procedures. Mahle and colleagues⁸ at the Children's Hospital of Philadelphia reported an improvement in hospital survival from 56.2% from 1984 through 1989 to 71.3% from 1995 through 1998 and 77.4% during 1998. At The Hospital for Sick Children in Toronto, hospital survival improved from 41% from 1990 through 1993 to 61% from 1994 through 1997 and 81% from 1998 through 2000.¹⁶ Hospital survival from 1992 through 1996 at the Children's Hospital of Wisconsin was 53%, whereas it reached 93% from 1996 through 2001.²¹ We have likewise experienced significant improvements at

the University of Michigan since first performing the Norwood procedure for HLHS in 1983. Meliones and coworkers²⁷ reported a 46% thirty-day survival for patients operated on between 1983 and 1989. From 1990 through 1997, hospital survival had improved to 76%, as reported by Lloyd and Bove.^{7,22}

In this analysis we found that many of the previously reported risk factors related to cardiovascular anatomy appear to have been minimized in the current era. The most commonly reported factors, including obstructed pulmonary venous return, ascending aortic diameter, anatomic subtype of HLHS, age at initial operation, preoperative pH, CPB time or operative time, and fetal diagnosis, could not be shown to have a significant effect on survival. Perhaps most surprising is the fact that obstructed pulmonary venous return did not reach significance. The most obvious explanation would be the limited sample size. However, recent advances in postoperative management, such as inhaled nitric oxide and sildenafil, might also have an effect. Perhaps most importantly, the objective of this study was to evaluate risk factors for hospital survival in the current era. The failure to define obstructed pulmonary venous return as a risk factor for hospital survival should not be misinterpreted to mean that it does not have a significant effect on the ultimate outcome of the patient with FSV. Underscoring

TABLE 4. Univariable analysis

	Survivor	Mortality	P value
Noncardiac anomaly	9% (8/88)	35% (8/23)	.0018
Weight <2.5 kg	5% (4/88)	17% (4/23)	.0072
HLHS with CAVV (unbalanced AVSD)	6% (5/88)	22% (5/23)	.017
Gestational age (wk)	40.1 (35-45.6)	39.4 (33.3-44.6)	.03
Weight (kg)	3.3 (1.9-4.5)	3.0 (1.3-4.5)	.04

HLHS, Hypoplastic left heart syndrome; CAVV, common atrioventricular valve; AVSD, atrioventricular septal defect.

TABLE 5. Multivariable analysis

	Survivor	Mortality	P value	Odds ratio
Weight <2.5 kg	5% (4/88)	17% (4/23)	.0098	7.0
Noncardiac anomaly	9% (8/88)	35% (8/23)	.032	6.0
HLHS with CAVV (unbalanced AVSD)	6% (5/88)	22% (5/23)	.083	3.7

HLHS, Hypoplastic left heart syndrome; CAVV, common atrioventricular valve; AVSD, atrioventricular septal defect.

this point, of the 7 hospital survivors, there was 1 late death of undetermined cause, 1 patient whose Fontan procedure was delayed because of marginal pulmonary vascular resistance, and 1 patient who is not a Fontan candidate and will ultimately require a cardiac transplantation because of pulmonary vascular obstructive disease.

Weight of less than 2.5 kg and noncardiac diagnoses continue to be significant risk factors for hospital survival for patients with HLHS or other FSV conditions. Although noncardiac anomalies might be more difficult to affect, recent advances might improve the survivals for the low-birth-weight FSV patient. The management of pulmonary blood flow is problematic in these smallest of patients. The traditional 3.5-mm modified Blalock-Taussig shunt results in pulmonary overcirculation and systemic malperfusion. The 3.0-mm graft leaves little room for technical error at the anastomoses and is potentially at increased risk of thrombosis. The recent renewed interest in the right ventricle-to-pulmonary artery conduit for the Norwood procedure might be a solution for these low-birth-weight patients. Although challenges remain, ongoing advances in preoperative, intraoperative, and postoperative management continue to improve the outlook for the patient with FSV.

Although this is a relatively large cohort of patients, it is possible that other risk factors might have been identified with a larger sample size. However, it is important to note that several of the previously identified risk factors have been identified from small groups of patients, suggesting that at a minimum, the effect of many of these risk factors has been lessened.

An analysis of the effect of DHCA versus RCP could not be performed. Data are listed and analyzed as total DHCA or RCP time. The study subjects are also participating in an ongoing randomized, prospective, evaluator-blinded trial. Because there were personnel common to both this article and to the blinded portion of the study, we were unable to perform this analysis without compromising the randomized trial.

This cohort is a heterogeneous population of patients undergoing a common operation. However, the numbers of patients are too small to perform meaningful subgroup analysis, which leaves the possibility that certain risk factors might apply to individual diagnostic groups.

The primary outcome for this study was hospital survival, and the follow-up is of an intermediate length. Patient characteristics, which might act as risk factors for late survival, cannot be defined.

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Appendix 1.

Comprehensive list of factors analyzed

Demographics	Preoperative factors
Age	Fetal diagnosis
Weight	History of cardiopulmonary arrest
Gestational age at birth	Lowest preoperative pH
Gestational age at operation	Immediate preoperative pH
Sex	Peak preoperative creatinine
Race	Immediate preoperative creatinine
Primary cardiac diagnosis	History of seizures
Other cardiac diagnoses	Obstructed pulmonary venous return
Other noncardiac diagnoses	Cause of obstructed pulmonary venous return
Anatomic subtype of hypoplastic left heart syndrome (aortic stenosis-atresia, mitral stenosis-atresia)	Need for balloon atrial septostomy
Morphologic right versus left ventricle	Echocardiographic data
Dysmorphic by physical examination	Aortic valve diameter and Z value
Chromosomal abnormality	Ascending aortic (at sinotubular junction) diameter and Z value
Genetic syndrome	Midtransverse arch diameter and Z value
	Mitral valve diameter and Z value
	Tricuspid valve diameter and Z value
	Ventricular function
	Antegrade aortic flow
	Degree of tricuspid regurgitation
	Degree of mitral regurgitation
	Degree of aortic regurgitation
	Degree of pulmonary regurgitation
	Degree of tricuspid regurgitation
	Operative data
	Total operative time
	Cardiopulmonary bypass time
	Deep hypothermic circulatory arrest or region cerebral perfusion time
	Shunt type
	Shunt size
	Hospital course
	Time to chest closure
	Time to initial extubation
	Days to initial intensive care unit discharge
	Days to hospital discharge
	Hospital survival
	Follow-up
	Length of follow-up
	Progression to hemi-Fontan procedure
	Progression to Fontan procedure
	Dead-alive
	Cause of death