

Home surveillance program prevents interstage mortality after the Norwood procedure

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Objective: To determine whether early identification of physiologic variances associated with interstage death would reduce mortality, we developed a home surveillance program.

Methods: Patients discharged before initiation of home surveillance (group A, n = 63) were compared with patients discharged with an infant scale and pulse oximeter (group B, n = 24). Parents maintained a daily log of weight and arterial oxygen saturation according to pulse oximetry and were instructed to contact their physician in case of an arterial oxygen saturation less than 70% according to pulse oximetry, an acute weight loss of more than 30 g in 24 hours, or failure to gain at least 20 g during a 3-day period.

Results: Interstage mortality among infants surviving to discharge was 15.8% (n = 9/57) in group A and 0% (n = 0/24) in group B ($P = .039$). Surveillance criteria were breached for 13 of 24 group B patients: 12 patients with decreased arterial oxygen saturation according to pulse oximetry with or without poor weight gain and 1 patient with poor weight gain alone. These 13 patients underwent bidirectional superior cavopulmonary connection (stage 2 palliation) at an earlier age, 3.7 ± 1.1 months of age versus 5.2 ± 2.0 months for patients with an uncomplicated interstage course ($P = .028$). A growth curve was generated and showed reduced growth velocity between 4 and 5 months of age, with a plateau in growth beyond 5 months of age.

Conclusion: Daily home surveillance of arterial oxygen saturation according to pulse oximetry and weight selected patients at increased risk of interstage death, permitting timely intervention, primarily with early stage 2 palliation, and was associated with improved interstage survival. Diminished growth identified 4 to 5 months after the Norwood procedure brings into question the value of delaying stage 2 palliation beyond 5 months of age.

Despite improved early outcomes after stage 1 palliation (S1P) for hypoplastic left heart syndrome (HLHS), there continues to be a 10% to 15% incidence of late death before stage 2 palliation (S2P), creation of the bidirectional cavopulmonary connection.^{1,2} Recurrent, residual, or progressive lesions after S1P that have been linked to interstage death include restrictive atrial septal defect, neo-aortic arch obstruction, systemic to pulmonary artery shunt stenosis, pulmonary artery distortion, coronary insufficiency, and atrioventricular valve insufficiency.³ Despite modifications of surgical technique and medical interventions, significant interim mortality continues to be associated with these anatomic lesions, which lead to progressive hypoxemia and impaired myocardial performance. In addition, hypovolemia and worsening hypoxemia related to common childhood illnesses are also associated with mortality among infants after S1P.⁴ To determine whether early identification of physiologic variances related to either anatomic changes or medical disease would reduce interstage mortality, we developed a home surveillance program for this at-risk population by enlisting parents to monitor daily arterial oxygen saturation via pulse oximetry (SpO_2) and weight at home between S1P and S2P.

Patients and Methods

Patients

Children's Hospital of Wisconsin institutional review board approval was obtained to assess the efficacy of a home surveillance program. Cohorts were created from all patients ($n = 87$) who underwent S1P for HLHS or a variant of HLHS at Children's Hospital of Wisconsin from July 1996 to November 2001. There were 63 patients in group A (July 1996 through September 2000), of whom 57 survived to hospital discharge after S1P and served as control subjects before initiation of the home surveillance program. The intervention group, group B (September 2000 through November 2001), included 24 patients, all of whom survived to hospital discharge after S1P and participated in the home surveillance program until returning for S2P.

Discharge Criteria

All patients were monitored with continuous pulse oximetry before discharge, with a target SpO_2 of greater than 75% both awake and asleep. If necessary, patients were given oxygen to achieve this goal and discharged home with supplemental oxygen. If unable to take in a full diet (110-130 kcal/[kg · d]) orally, a gastrostomy tube was placed to provide caloric supplementation; 1 patient was discharged home with a nasogastric tube in place. Digoxin was prescribed for all patients. Afterload reduction with captopril and diuretic therapy with furosemide were initiated with weaning of inotropic support and used selectively for those patients demonstrating an elevated pulmonary to systemic flow ratio greater than 2.0 ($Q_p/Q_s > 2.0$) in the early postoperative period. Medical therapy was adjusted on an outpatient basis at the discretion of the treating cardiologist. Cardiologists practicing at Children's Hospi-

tal of Wisconsin followed up 93% of patients in group A and 67% of patients in group B.

Home Management

Parents of group A patients received the usual discharge instructions, including directives to contact their children's physician for respiratory or gastrointestinal illness, respiratory difficulties (tachypnea, accessory muscle use, nasal flaring, distress), and changes in perfusion (cool extremities, dusky or ashen appearance). Other than scheduled follow-up with the cardiologist every 2 to 4 weeks, no additional home surveillance was performed in group A. In addition to the usual discharge instructions, patients in group B were sent home with digital infant scales sensitive to changes of 10 g (Baby Checker Scale; Medela, McHenry, Ill) and pulse oximeters (Nellcor 200; Nellcor Puritan Bennett Inc, Pleasanton, Calif). Parents were asked to record weight and SpO_2 in a daily log. Thresholds for parents to seek medical advice included resting SpO_2 less than 70%, weight loss of 30 g or failure to gain 20 g of weight during 3 days. Patients who breached surveillance criteria underwent physician evaluation within 24 hours.

Data Analysis

A prospective perioperative database containing demographic, surgical, hemodynamic, laboratory, and nutritional data for all patients who underwent S1P was reviewed. Patient-related variables assessed included age, sex, weights at birth and at S1P, diagnostic category, ascending aortic diameter, need for preoperative mechanical ventilation or inotropic support before S1P, and feeding method at time of hospital discharge. Operative variables included cardiopulmonary bypass time, deep hypothermic circulatory arrest time, total support time (defined as cardiopulmonary bypass time plus deep hypothermic circulatory arrest time), and shunt size. Hemodynamic variables compared during the first 48 postoperative hours included arterial and venous oxygen saturations, arteriovenous oxygen content difference, Q_p/Q_s , hemoglobin, mean blood pressure, central venous pressure, and anaerobic threshold. Variables assessed at the time of S2P included age, weight, and preoperative oxygen saturation. Outcome measures were operative survival after S1P and survival to S2P.

Interstage weights and SpO_2 were obtained at periodic clinic visits for patients who were discharged home without home surveillance (group A, $n = 57$) and were compared with prospectively collected data from patients discharged with home monitoring equipment (group B, $n = 24$). Interstage SpO_2 and weight were compared between the two groups. Fixed effects regression models were used to compare differences in SpO_2 between groups A and B, as well as within group B for comparison of patients who did versus did not undergo any intervention as a result of home monitoring.

Statistical analysis was completed with SPSS advanced Models 9.0 (SPSS, Inc, Chicago, Ill) and STATA software (Stata Corporation, College Station, Tex). Descriptive statistics are presented as mean \pm SD or percentage and count unless otherwise indicated. Median values are reported where appropriate. Variables were analyzed with χ^2 statistics for categorical variables and analysis of variance techniques for continuous variables. Actuarial survival analysis was performed with Kaplan-Meier methods with log-rank comparison of cumulative survival by group. A polynomial regres-

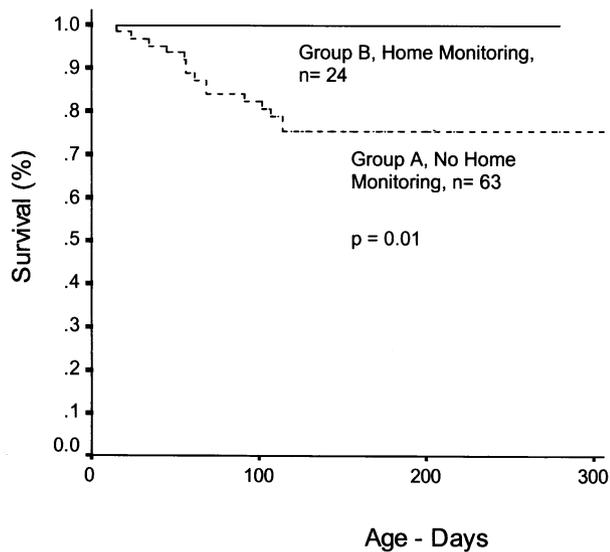


Figure 1. Survival curve calculated by Kaplan-Meier method for patients between S1P and S2P. Interstage survival was significantly improved in group B.

sion equation was generated to plot patient growth between S1P and S2P (SPSS).

Results

Follow-up data were available for 100% of the patients through May 2002. Early survival after S1P was 90.4% (57/63) for group A and 100% (24/24) for group B. Interstage mortality among survivors to hospital discharge before S2P was 15.8% (9/57) in group A and 0% (0/24) in group B ($P = .039$). Survival from birth through S2P was 74.6% (48/63) in group A and 100% (24/24) in group B ($P = .01$; Figure 1). In the home surveillance group (group B), the age at S2P was younger, 4.3 ± 1.6 months compared with 5.6 ± 2.1 months in group A ($P = .016$). Thirteen of 24 patients in the home surveillance group (group B) breached home monitoring criteria. These 13 group B patients underwent earlier S2P (3.7 ± 1.1 months of age) than the remaining 11 patients (5.2 ± 2.0 months of age; $P = .028$). The 11 group B patients who did not breach surveillance criteria underwent S2P at an age that was not different from that of patients in group A (5.2 ± 2.0 months vs 5.6 ± 2.1 months). There was 1 in-hospital death among patients undergoing S2P in group A; there were no in-hospital deaths in group B.

Patient and Operative Variables

The anatomic diagnoses did not differ between groups and are shown in Table 1. During this study period, no patient was deemed to be ineligible for or denied S1P, and no patient underwent heart transplant as primary therapy. S1P was declined by the parents of one neonate with HLHS and multiple congenital anomalies. Five other patients with ex-

TABLE 1. Anatomic diagnoses, group A versus group B

Diagnostic category	Group A (n = 63)	Group B (n = 24)
HLHS	45 (71.5%)	19 (79.2%)
Aortic atresia with mitral atresia or mitral stenosis	35	15
Aortic stenosis with mitral atresia or mitral stenosis	10	4
Hypoplastic left heart variant	18 (28.5%)	5 (20.9%)
HLHS with ventricular septal defect	7	1
Heterotaxy	3	0
Unbalanced atrioventricular canal	1	1
Complex double-outlet right ventricle	3	0
Tricuspid atresia with transposition of the great arteries	1	1
Double-inlet left ventricle with transposition of the great arteries	3	2

Differences between groups in proportions of HLHS and hypoplastic left heart variant diagnosis were not significant.

tracardiac anomalies, all from group A, underwent successful S1P, with 4 of 5 patients surviving to S2P. A patient with Turner syndrome died before S2P. The extracardiac anomalies of the interstage survivors included tracheoesophageal fistula with foramen of Morgagni diaphragmatic hernia, cystic fibrosis, ciliary dysmotility, and chromosome 15 long-arm deletion. Aortic atresia with mitral atresia or stenosis was the predominant diagnosis in each group of patients discharged to home after S1P: 33 of 57 (61.4%) in group A and 15 of 24 (62.5%) in group B. There was a trend toward smaller ascending aortic diameter in group B patients (2.67 ± 0.99 mm vs 3.42 ± 1.67 mm, $P = .06$). The finding of ascending aortic diameter less than or equal to 2 mm was not different between groups: 40% in group A and 55% in group B. Prenatal diagnosis of HLHS was increasingly common in the more recent cohort (62% in group B vs 19% in group A; $P < .001$). The groups did not differ in gender, need for preoperative mechanical ventilation or inotropic support, or age or weight at the time of S1P. Cardiopulmonary bypass time was significantly longer in group B (125.4 ± 48.0 minutes vs 171.1 ± 47.1 minutes, $P < .001$), with correspondingly shorter circulatory arrest time (59.8 ± 17.0 minutes vs 8.3 ± 3.0 minutes, $P < .001$; Table 2). The differences in cardiopulmonary bypass and circulatory arrest times between groups reflect the use of continuous cerebral perfusion in group B. However, total support times did not differ between groups (183.3 ± 56.0 minutes vs 179 ± 47.3 minutes). Postoperative arterial and venous oxygen saturations, arteriovenous oxygen content

TABLE 2. Patient and operative characteristics at S1P, group A versus group B

Variable	Group A (n = 63)	Group B (n = 24)	P value
Gender (% male)	65%	54%	NS
Age at S1P (d)	8.1 ± 9.0	6.4 ± 4.9	NS
Weight at S1P (kg)	3.17 ± 0.56	3.15 ± 0.34	NS
Aortic diameter (mm)	3.42 ± 1.67	2.67 ± 0.99	.06
Cardiopulmonary bypass time (min)	125.4 ± 48.0	171.1 ± 47.1	<.001
Circulatory arrest time (min)	59.8 ± 17.1	8.3 ± 3.0	<.001
Total support time* (min)	185.3 ± 56.0	179.4 ± 47.3	NS
Median shunt size (mm)	3.5	3.5	NS
Hospital stay (d)	35 ± 19	31 ± 13	NS
Early survival (%)	90.4%	100%	NS

Values are mean ± SD unless otherwise indicated. NS, Not statistically significant.

*Cardiopulmonary bypass time plus deep hypothermic circulatory arrest time.

difference, \dot{Q}_p/\dot{Q}_s , hemoglobin, mean blood pressure, central venous pressure, and anaerobic threshold did not differ between groups during the first 48 hours.

Outpatient Variables

Atrioventricular valve insufficiency (moderate or greater) was present at hospital discharge in 17.5% of patients in group A and 25% of patients in group B. Ten patients in group A (17.5%) were discharged home with supplemental oxygen, as were 4 patients in group B (16.7%). Mean SpO_2 values at discharge were $81.1\% \pm 4.2\%$ in group A and $82.4\% \pm 2.8\%$ in group B. At the time of S2P, SpO_2 values were $74.2\% \pm 6.0\%$ in group A and $72.5\% \pm 5.4\%$ in group B. Within group B, patients who underwent an intervention as a result of increased cyanosis detected at home were more hypoxemic at S2P than were those patients who did not have increased cyanosis detected at home (SpO_2 $75.8\% \pm 3.5\%$ vs $69.8\% \pm 5.4\%$, $P = .005$; Table 3). Figure 2 illustrates the decrease in SpO_2 that prompted intervention in the home surveillance group relative to the SpO_2 trend in group B patients who had no home-detected events ($P < .001$ by fixed effects regression).

Weight at hospital discharge after S1P was not different between groups: 3.35 ± 0.63 kg in group A versus 3.21 ± 0.40 kg in group B. Gastrostomy tubes were placed in 28% of the patients in group A and 25% of patients in group B. Despite younger age at S2P for group B, weight at the time of S2P was not different between groups (5.70 ± 1.35 kg in group A vs 5.34 ± 0.80 kg in group B; Table 3). From more

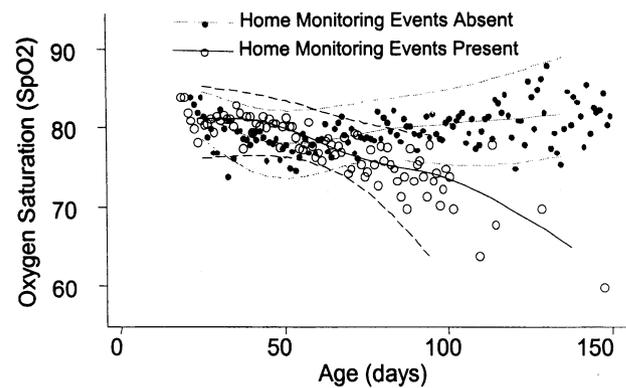


Figure 2. SpO_2 trends for group B. Locally weighted polynomial regression lines and 95% confidence interval are indicated. This illustrates that patients in group B who underwent intervention because of home-detected events had decline in SpO_2 not seen in patients without events detected at home and who did not require early intervention. Patients who had events detected had significantly lower SpO_2 than those who did not ($P < .001$ by fixed exact regression).

than 1400 patient weights obtained for the entire cohort, a polynomial regression equation was generated to estimate patient growth between S1P and S2P (Figure 3). Average daily weight gain of the cohort was 20 g/d. The growth curve generated demonstrated a slowing of growth velocity between 4 and 5 months of age.

Mortality and Interstage Interventions in Group A

Interstage mortality in group A was 15.8% (9/57), with all deaths occurring at less than 120 days of age (Figure 4). Autopsy data were available for 3 of 9 patients. Clinical impressions and echocardiographic findings before and at the time of death were obtained through chart reviews for all 9 patients. Aortic atresia was the diagnosis in 78% of the interim death group (7/9). Interstage death was associated with residual or recurrent lesions, concurrent illness, or feeding difficulties. Some patients had more than one problem. One patient with aortic atresia, mitral stenosis, and Turner syndrome underwent balloon dilatation of distal shunt obstruction before discharge after S1P. She subsequently had progressive tricuspid insufficiency and ventricular dysfunction develop. This was the only patient with a residual lesion identified before hospital discharge. Two other patients, both with aortic and mitral atresia, had outpatient echocardiographic findings of narrowing at the distal end of the shunt and moderate tricuspid regurgitation. One of these patients also had concomitant neo-aortic coarctation gradient of 60 mm Hg. Among the remaining 6 patients, we did not identify shunt obstruction, arch obstruction, or compromise of the atrial septal defect. Two patients with no prodromal illness died suddenly at home. One of these patients was known to have moderate tricuspid regurgita-

TABLE 3. Patient characteristics at S1P discharge and S2P

Variable	Group A (n = 57)	Group B (n = 24)	P value	B ₀ (n = 11)	B ₁ (n = 13)	P value
Gastric tube placement (%)	28%	25%	NS	27%	23%	NS
S1P discharge weight (kg)	3.4 ± 0.6	3.2 ± 0.4	NS	3.1 ± 0.4	3.3 ± 0.4	NS
Weight at S2P (kg)	5.7 ± 1.3	5.3 ± 0.8	NS	5.4 ± 0.8	5.3 ± 0.8	NS
Home oxygen therapy	17.5%	16.7%	NS	8.3%	8.3%	NS
SpO ₂ at discharge (%)	81.1 ± 4.2	82.4 ± 2.8	NS	82.8 ± 2.1	82.2 ± 3.5	NS
SpO ₂ at S2P (%)	74.2 ± 6.0	72.5 ± 5.4	NS	75.8 ± 3.5	69.8 ± 5.4	.005
Age at S2P (mos)	5.6 ± 2.1	4.3 ± 1.6	.016	5.2 ± 2.0	3.7 ± 1.1	.028
Survival to S2P	76%	100%	.01	100%	100%	NS
Hospital stay for S2P (d)						NS
Mean	13 ± 13	13 ± 21	NS	16 ± 25	9 ± 3	
Median	8	8		7	8	

Comparisons are group A versus group B and within group B between patients without an intervention as a result of home surveillance (B₀) versus those with an intervention (B₁). Values are mean ± SD unless otherwise indicated. NS, Not statistically significant.

tion. Three patients had evidence of a respiratory illness at the time of death. Two of these patients had respiratory infection confirmed at autopsy, 1 with unspecified viral pneumonia and 1 with *Pneumocystis carinii* pneumonia. Four patients had feeding difficulties. One patient, with a several day history of decreased oral intake, died suddenly a week after diuretic therapy was increased. One patient in group A was seen at 111 days of age critically ill and in shock with decreased SpO₂ after a 2-day history of diarrhea. This patient underwent emergency S2P for what was thought to be impending shunt thrombosis. This patient was noted to have ischemic necrosis of the bowel in the operating room at the end of the procedure. Bowel ischemia confirmed by pathologic examination was determined to have predated the S2P procedure and was not due to embolism or thrombosis of the superior mesenteric artery but rather was consistent with hypovolemia. This was the only in-hospital death among any of the patients undergoing S2P. Although this was not strictly an interstage death, the mechanism of death was consistent with those observed in the interstage mortality group.

Interventions for recurrent arch obstruction were performed in 33% of the patients in group A (19/57). Interventional catheterization with balloon angioplasty for recurrent arch obstruction was performed in 3 of 57 (5.3%). Surgical arch revision was performed in 3 of 57 patients (5.3%) before S2P, whereas 13 of 57 (22.8%) underwent arch revision at the time of S2P. None of the patients who underwent balloon angioplasty had subsequent surgical arch revision at S2P.

Mortality and Interstage Interventions in Group B

All patients discharged with home monitoring of SpO₂ and weight survived to S2P. Data obtained through home surveillance found 13 of 24 patients at increased risk: 12 patients with worsening hypoxemia from baseline and 1

patient with poor feeding and poor growth without worsening hypoxemia.

Outpatient detection of desaturation from baseline, as illustrated in Figure 2, was the predominant cause for intervention in group B and occurred 14 times in 12 of 24 home surveillance subjects. Parents of all but 3 patients sought medical attention at age less than 100 days (range 41-99 days, median 69 days) as the result of breach of home surveillance criteria. Two of 12 patients were hospitalized at 41 and 88 days of age with cough, congestion, hypoxemia, and need for frequent nasopharyngeal suctioning, consistent with bronchiolitis (1 had a positive culture for respiratory syncytial virus). These 2 patients each were seen a second time by the cardiologist for increased cyanosis without an associated respiratory illness, which led to a diagnostic cardiac catheterization and subsequent S2P at the ages of 113 and 108 days. Of the remaining 10 patients with increased interstage cyanosis detected at home, all underwent cardiac catheterization within 7 days of presentation (median age 78 days, range 59-182 days). Eight patients underwent catheterization at less than 100 days of age. Seven patients underwent subsequent S2P within 3 days of catheterization, and 1 patient underwent balloon angioplasty of the systemic to pulmonary artery shunt at 69 days of age with subsequent S2P performed at 129 days of age because of poor growth. Among the 7 patients who had subsequent S2P within 3 days of catheterization, 2 patients were found to have narrowing of the origin of the innominate artery from which the systemic to pulmonary shunt arose and 3 patients had distal shunt narrowing. Two patients without residual or recurrent anatomic lesions and without obstruction of shunt flow were thought to be favorable candidates for early S2P and underwent subsequent palliation at ages 79 and 90 days of age because of unacceptable cyanosis. Another of the 10 patients with increased interstage cyanosis was not found to have a recurrent or residual anatomic

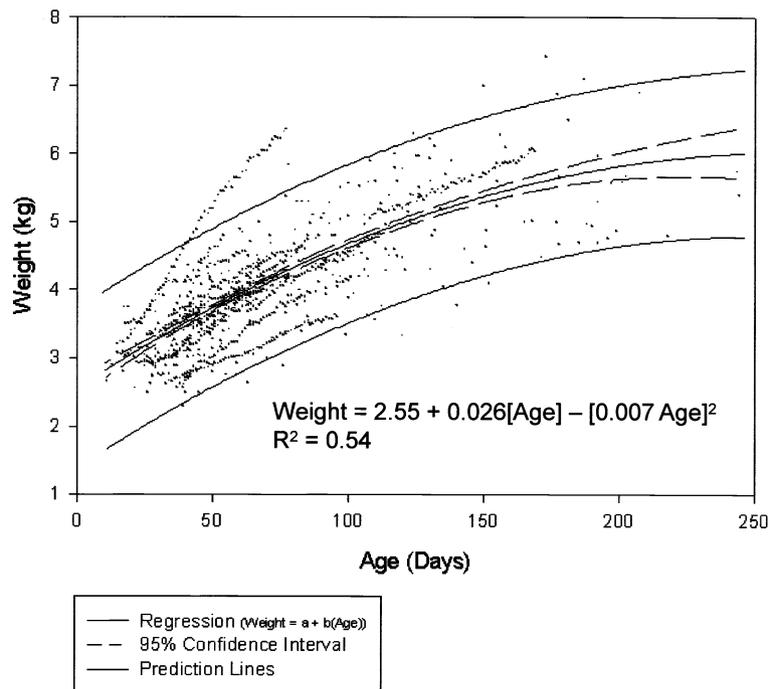


Figure 3. Interstage patient growth among survivors to S2P was calculated with more than 1400 observations of patient weight between S1P and S2P. Regression line and 95% confidence intervals are indicated. Curve indicates limited growth potential after S1P of HLHS. In this group of patients, unlike healthy infants in whom continued growth is expected, growth into later infancy was limited. This is further evidence of interstage period risk.

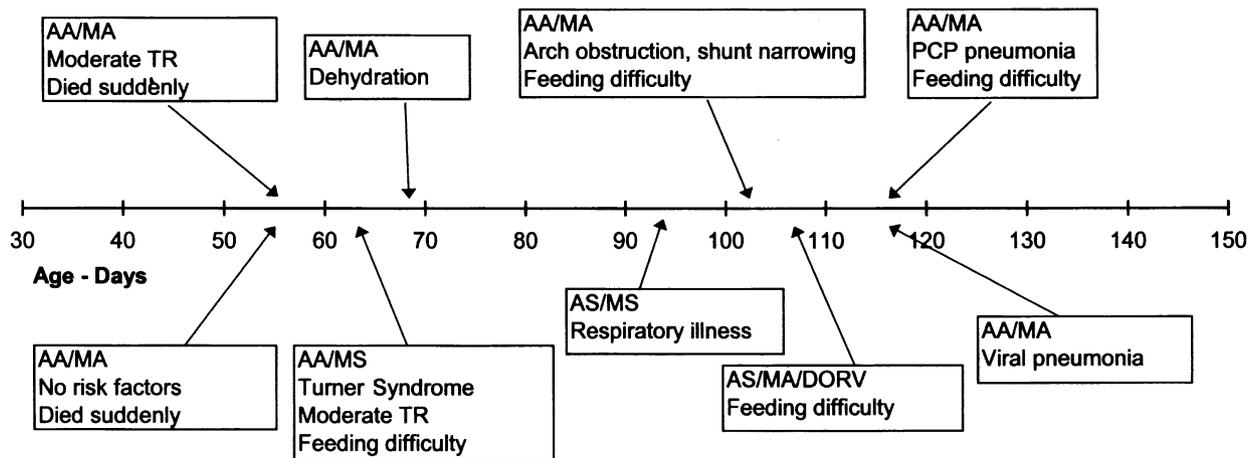


Figure 4. Timeline of interstage mortality and risk factors for group A. Only one patient died suddenly without residual or recurrent lesions or antecedent illness. Tricuspid insufficiency indicates insufficiency graded as moderate or greater in severity. AA, Aortic atresia; MA, mitral atresia; TR, tricuspid regurgitation; PCP, *P carinii* pneumonia; MS, mitral stenosis; AS, aortic stenosis; MA, mitral atresia; DORV, double-outlet right ventricle.

lesion at 59 days of age but was responsive to oxygen. He was thought to have medical lung disease and was discharged home with supplemental oxygen to undergo S2P 2.5 months later. One patient who had an elective preoper-

ative diagnostic catheterization was admitted 2 weeks before scheduled S2P for earlier intervention related to worsening hypoxemia. In 8 of 24 home surveillance patients, interval hypoxemia led to S2P at less than 100 days of age.

Weight loss criteria were not violated by any patient, nor was any patient noted to have gastrointestinal losses. Of the 12 patients with increased interstage cyanosis, 4 (all less than 100 days old at 55, 69, 76, and 92 days) also had decreased feeding and poor weight gain reported. The patient who was seen at 55 days of age with poor feeding underwent gastrostomy tube placement for nutritional support. One patient with poor feeding and poor growth who did not have concomitant desaturations from baseline underwent cardiac catheterization followed by S2P with arch reconstruction at 113 days of age. No patient in group B underwent interventional catheterization for recurrent arch obstruction or surgical arch reconstruction before S2P; however, 6 of 24 patients (25%) underwent arch reconstruction at the time of S2P.

Discussion

The effectiveness of staged single-ventricle palliation for HLHS has been limited by high mortality. Improved postoperative management that focuses on control of the \dot{Q}_p/\dot{Q}_s combined with early detection of inadequate perfusion has resulted in improved S1P survival.⁵⁻⁷ Mortality before S2P remains high, and it is unclear whether improvements in S1P management have affected this high-risk interstage period.^{1,2} Even the optimal S1P patient continues to have physiologic risks, specifically parallel circulation, volume overload to the single ventricle, and cyanosis. The introduction of S2P as an intermediate step in single-ventricle palliation was a milestone that resulted in improved survival and allowed the practical application of the Fontan pathway to patients with HLHS. The benefits of S2P include relief of excess volume load and improvement in arterial saturation. The S2P procedure is well tolerated in this group of patients, with low mortality and short hospital stays. After S2P, patients are at substantially decreased risk of death, show improved growth, have better tolerance of concurrent illness, and are low-risk candidates for a completion Fontan operation.⁸⁻¹¹ The optimal timing of S2P has not been determined. Although patients younger than 4 weeks tolerate S2P poorly, infants as young as 8 weeks with satisfactory preoperative hemodynamics have undergone S2P successfully.¹² Earlier relief of volume overload and improvement in cyanosis may be especially important in the patient with a single right ventricle with limited ability to increase cardiac output and a tricuspid valve prone to development of insufficiency when exposed to prolonged volume or pressure overload.

During the interstage period, patients are at risk for development of recurrent lesions, such as shunt stenosis or arch obstruction, that may be subtle and have important consequences. Jonas³ has previously described restrictive atrial septal defect, neo-aortic arch obstruction, pulmonary artery distortion, and tricuspid valve insufficiency as ana-

tomically abnormalities that may contribute to attrition after successful S1P. In a series of postmortem evaluations, Bartram and colleagues¹³ confirmed these findings, noting residual or recurrent lesions in 44 of 122 deaths (36%).

Any process that causes an imbalance in the oxygen supply-demand relationship will result in decreased arterial oxygen saturation in the patient with parallel circulation. An example of this is respiratory infection, which may result in pulmonary venous desaturation and worsening cyanosis because of impaired respiratory mechanics resulting from secretions, airway edema, and pneumonitis. This would support previous reports of viral illnesses as a cause of death in patients after S1P.^{4,13,14} In this study, of the 9 interstage deaths before implementing the home surveillance program, 4 (44%) patients had symptoms of a respiratory illness or dehydration. Gastroenteritis, a common illness in infants, may result in acute dehydration. In the patient with single-ventricle anatomy after S1P, the resulting increase in sympathetic tone will lead to further reduction of systemic flow as \dot{Q}_p/\dot{Q}_s increases. One additional patient who died after emergency S2P had evidence of a preoperative gastrointestinal pathologic condition that might have been identified through acute weight loss. Indeed, it was this patient who motivated us to develop a home surveillance program.

The primary goal of the home surveillance program was to develop a simple, reliable strategy to detect worsening systemic oxygenation and acute dehydration. Patients were discharged with infant scales, to identify dehydration through acute weight loss as well as growth failure through lack of weight gain, and with pulse oximeters, to identify worsening desaturation. These were devices with which the parents were already familiar and that were straightforward to use. The criteria for contacting a physician were determined by consensus as representing the physiologic limits beyond which survival would be in jeopardy and included SpO_2 less than 70% and acute weight loss of 30 g or failure to gain 20 g during 3 days. Worsening desaturation was the most common indication for contacting a physician and occurred in 50% of the patients (12/24). No patient had acute dehydration or the clinical appearance of gastroenteritis, but isolated poor weight gain resulted in a gastrostomy tube in 1 patient and early S2P with repair of recurrent arch obstruction in another. Overall timing of S2P was earlier in group B than group A (5.6 ± 2.1 months vs 4.3 ± 1.6 months, $P = .016$). Of significant interest, though, is the intragroup analysis of group B patients. In 13 of 24 patients with evidence of increased vulnerability (54%), as detected by decreased SpO_2 or slowed weight gain, progression to S2P occurred significantly earlier than in the remaining 11 patients in group B who had no detectable problems at home (3.7 ± 1.1 months vs 5.2 ± 2.0 months, $P = .028$). Additionally, the age at S2P of group B patients who did not breach surveillance criteria was not different from the age of

S2P for group A patients (5.2 ± 2.0 months vs 5.6 ± 2.1 months).

A growth curve that included more than 1400 data points was developed for all the survivors to S2P. Unlike the growth curve of a healthy infant, who usually doubles the birth weight by 5 months of age, the patient with HLHS who has undergone S1P appears to have limited growth potential, with a plateau phase of weight gain after 150 days (Figure 3). This limited growth potential provides further evidence of the increased risk of the interstage period. Interestingly, although group A patients were older at the time of the S2P, they weighed the same as the patients in group B. The group A patients may have been subjected to a period of prolonged cyanosis and volume overload without the benefit or perhaps even the possibility of additional weight gain. This poor growth potential after 4 to 5 months calls into question the value of routinely delaying S2P beyond 5 months of age.

Previous studies have identified aortic atresia and smaller ascending aortic diameter as risk factors for late death after S1P.¹⁵⁻¹⁷ This anatomic subtype represents the most extreme form of HLHS, presumably with the lowest physiologic reserve. There were no differences in anatomic subtypes between groups, although there was a trend toward smaller ascending aortic size in the home surveillance group. Thus the survival advantage in group B was not based on favorable anatomy.

The data suggest that frequent monitoring of SpO_2 and weight were useful in selecting patients at increased risk during the interstage period. In the patient with single-ventricle anatomy and parallel circulation, arterial saturation is a function, among other things, of hemoglobin, pulmonary venous saturation, \dot{Q}_p/\dot{Q}_s and total cardiac output. Therefore, diminished SpO_2 may be particularly sensitive and will discriminate patients with anemia, respiratory infection, decreased total cardiac output, as well as decreased \dot{Q}_p/\dot{Q}_s . It is more difficult to develop a home surveillance strategy to reliably detect recurrent arch obstruction. Recurrent arch obstruction, however, might be identified through the more subtle symptom of slow weight gain.

The home surveillance program was associated with improved interstage survival; however, this study has important limitations. This study compared noncontemporary groups, although it should be noted that preoperative and postoperative management was uniform and postoperative hemodynamics did not differ between groups. Modification of surgical technique, more specifically implementation of continuous cerebral perfusion, resulted in shorter circulatory arrest time in the more recent cohort and may be implicated as a variable that improved interstage survival. We cannot rule out the impact of prenatal diagnosis on interstage survival. This study was neither blinded nor ran-

domized, and therefore we cannot definitively conclude that lack of intervention in patients who breached surveillance criteria would have resulted in death. Overall progression along an institutional learning curve also probably contributed to improved outcomes in the home surveillance group. Despite these limitations, criteria selected to prompt examination were not different from those that would have triggered investigation in either group if identified at a routine clinic visit. In addition, it is important to note that the development of desaturation and poor weight gain occurred abruptly during the course of several days, a shorter interval than even reasonably spaced clinic visits.

The success of a home surveillance program requires dedicated family participation, as well as collaboration among multidisciplinary health care providers, which may be challenging for a given patient. Obtaining reliable data through appropriate equipment use is necessary for adequate assessment of physiologic variances and could prove to be a limitation. These patients frequently have a prolonged intensive care unit course, and parents become familiar with the concept and measurement of SpO_2 . We were impressed that parents came to identify episodes and recognize the significance of decreased saturation while their children were in the hospital. They appeared to be reassured when arterial saturations were in an acceptable range. The psychosocial impact of home SpO_2 and weight monitoring was not formally evaluated. Given the risk of interstage death, however, we suspect that families have taken comfort in having objective data as an indicator of their children's condition.

Improvements in S1P operative survival were, in our experience, the result of objective assessment of the patients' circulatory status through improved physiologic monitoring. In this study improvement in interstage survival was also the result of continued collection of objective data through home surveillance to select patients at risk for interstage death. In this small series, we discriminated patients who were at increased risk for interstage death because of concurrent illness or residual or recurrent lesions. Early S2P was the primary strategy used to treat this subgroup of patients who appeared to be in jeopardy during the interstage period. It is clear from our experience and those of others that although S2P can be successfully accomplished in patients as young as 6 weeks, the postoperative course in these patients is prolonged relative to older patients. We must assume that the very young patient is at increased risk for death after S2P. Thus proposing early S2P for everyone, rather than reserving this therapy for those in whom the risk-benefit ratio is favorable, may in fact compound morbidity and mortality. Because growth appears to plateau between 4 and 5 months of age, we can conclude that S2P should be completed no later than 5 months of age. Continued data collection and follow-up are necessary to

determine whether these short-term improvements will result in improved long-term outcomes. We continue to use home surveillance to discriminate patients at risk for interstage death.

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Discussion

Dr J. William Gaynor (Philadelphia, Pa). I congratulate Ghanayem and coauthors on this excellent report addressing the important and troubling issue of interstage mortality after successful S1P. This is yet another important contribution to the management of these difficult cases from your institution.

In this study, detection of home-monitored events, either decreased oxygen saturation or poor weight gain, led to earlier evaluation of these patients and frequently earlier performance of the superior cavopulmonary connection. There was no interstage mortality in the relatively small group of infants for whom home monitoring was used.

Dr Ghanayem, I have several questions. First, we recently reviewed risk factors for death after S1P in 158 infants during a 3.5-year period. In 46% of these infants there were additional risk factors, such as low birth weight (<2.5 kg); presence of additional cardiac defects at the time of S1P, such as interrupted aortic arch, moderate to severe tricuspid regurgitation, and anomalies of pulmonary or venous drainage; or presence of major extracardiac anomalies or genetic syndromes.

The interstage mortality in that series was approximately 10%, similar to your group A. In a multivariable analysis, the strongest predictors of interstage mortality were the presence of additional cardiac defects and the presence of an extracardiac anomaly or genetic syndrome. In your series, how many infants had either additional cardiac defects or extracardiac anomalies? And what was the impact of these associated anomalies on interstage mortality?

Second, it would appear that in group A several of the deaths occurred at a relatively early age, 50 to 70 days. These deaths appeared to occur before the age at which home monitoring events were detected in group B and before the age at which most group B patients underwent cavopulmonary connection. Do you attribute the lack of these early deaths and this decreased very early mortality to the home monitoring program, or does this simply represent increasing institutional experience with the management of these infants or some other change in management?

Third, an intriguing finding of your study is the decrease in growth velocity at approximately 5 months of age. This implies that there may be no advantage to delaying cavopulmonary connection even for children who are doing relatively well. What is your current policy for timing of cavopulmonary connection for children who have no home monitoring events and are otherwise doing well?

Fourth, the presence of aortic atresia, particularly with a diminutive ascending aorta, is a potential risk factor for coronary ischemia and interstage mortality. What was the incidence of very small ascending aorta, less than 2 mm, in your series? Was there any difference between group A and group B?

Finally, I was intrigued by the fact that many of your patients with recurrent arch obstruction undergo surgical intervention rather than balloon dilatation of the arch. What are your reasons for choosing surgical reconstruction rather than balloon dilatation?

I again congratulate you on your excellent study.

Dr Ghanayem. Thank you, Dr Gaynor, for your remarks. The presence of additional cardiac defects, such as preoperative moderate to severe tricuspid regurgitation, anomalous origin of the