Improving interstage survival after Norwood operation: Outcomes from 10 years of home monitoring

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Objective: Infants who undergo Norwood stage 1 palliation (S1P) continue with high-risk circulation until stage 2 palliation (S2P). Routine care during the interstage period is associated with 10% to 20% mortality. This report illustrates the sustained reduction of interstage mortality over 10 years associated with use of home monitoring.

Methods: Daily monitoring of oxygen saturation and weight was done for all patients discharged to home after S1P. Notification of the care team occurred for oxygen saturation <75% or >90%, weight gain < 20 g over 3 days, weight loss > 30 g, or intake < 100 cc/kg/d. Breach of these criteria marked an interstage event. Interstage outcomes are reported. Patient characteristics and perioperative variables were compared between patients with and without interstage events.

Results: Over 10 years, 157 patients were discharged after S1P with home monitoring. Interstage survival was 98%. Breach of home criteria occurred in 59% (93 out of 157), with violation of oxygen saturation <75% the most common event. Patient characteristics, operative data, and early postoperative morbidity did not differ between patients with and without events.

Conclusions: Home monitoring after S1P is associated with excellent interstage survival. Although a breach of monitoring criteria occurred in more than half of patients, our analysis failed to identify independent predictors of interstage events. Analysis of variables predicting mortality could not be assessed due to the low frequency of death in this cohort. Failure to identify specific variables for interstage events suggests that home monitoring, as part of an interstage surveillance program, should be applied to all S1P hospital survivors. (J Thorac Cardiovasc Surg 2014;148:1540-7)

Advances in surgical and perioperative care of infants undergoing Norwood stage 1 palliation (S1P) have resulted in 80% to 95% survival to hospital discharge.¹⁻⁵ The continued risk of death during the interstage period, following S1P hospital discharge and before stage 2 palliation (S2P) with the superior cavopulmonary connection, highlights the ongoing vulnerability inherent to single ventricle anomalies. Specifically, these infants are subject to systemic to pulmonary artery shunts, excessive volume loading on the systemic ventricle, and cyanosis. Several single-center studies report interstage mortality rates of 10% to 20%.^{4,6-11} Comparably, interstage mortality was 12% in the multicenter Pediatric

Heart Network Single Ventricle Reconstruction (SVR) trial of infants with hypoplastic left heart syndrome (HLHS) and single right ventricle variants.^{6,13}

Interstage mortality has been associated with various patient characteristics; anatomic risk factors, including residual lesions; surgical technique at initial palliation; postoperative complications; arrhythmias; and acquired intercurrent illness.^{4,6-12,14-17} Several of these factors can lead to excessive hypoxemia, hypovolemia, alteration in systemic vascular resistance, and/or progressive myocardial dysfunction. These derangements consequently place infants with parallel circulation and minimal myocardial reserve at risk for interstage death before S2P. Without the ability to definitively predict who is at risk for interstage death, home monitoring strategies have been implemented to detect physiologic variances that may precede clinical deterioration. Early experience with home monitoring of daily oxygen saturation levels and weight change has been associated with a marked reduction in interstage mortality after S1P and before S2P through timely recognition of physiologic vulnerability.^{7,9,18} Since our initial report of interstage surveillance for HLHS, the home monitoring program (HMP) at Children's Hospital of Wisconsin has undergone a series of modifications in

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Abbreviations and Acronyms

ECMO	= extracorporeal membrane oxygenator
HLHS	= hypoplastic left heart syndrome
HMP	= home monitoring program
mBTs	= modified Blalock-Taussig shunt
S1P	= stage 1 palliation
S2P	= stage 2 palliation
SpO ₂	= oxygen saturation
SVR	= Single Ventricle Reconstruction

response to ongoing quality improvement cycles in the face of continued interstage morbidity and mortality. This report illustrates the impact of 10 years of our HMP targeted at reducing interstage mortality in patients undergoing the Norwood operation.

METHODS

Patient Population

Since October 2000, all patients discharged to home following S1P at Children's Hospital of Wisconsin have been enrolled in the interstage HMP as a standard of clinical practice. All S1P survivors discharged to home before S2P comprised this study cohort. Patients who remained hospitalized to S2P were excluded from the primary analysis; however, their outcomes are reported here. Local institutional review board approval and parental consent was obtained for perioperative and interstage prospective data collection.

Discharge Criteria

Continuous pulse oximetry, daily weights, and fluid intake are monitored with set goals before hospital discharge. Target oxygen saturation (SpO₂) is >75% while awake and asleep. Patients with oxygen-responsive hypoxia that is not secondary to an anatomic lesion may be discharged to home on supplemental oxygen. All patients discharged must consume a minimum of 100 cc/kg/d and demonstrate weight gain before discharge. A caloric goal of 110 to 130 kcal/kg/d is generally needed for somatic growth; however, patients able to gain weight with fewer calories do not need to meet the specified caloric goal. Patients unable to consume sufficient enteral volume or calories with oral feeds have a surgical gastrostomy tube placed without Nissen fundoplication. Patients remained as inpatients throughout the interstage period if they had complex extracardiac medical needs or concerning circulatory instability determined by the cardiac critical care team.

In preparation for home monitoring, parents undergo a comprehensive education program before discharge that includes proper use of the infant scale and pulse oximeter, review and calculation of fluid volume and nutritional intake goals, calculation of incremental changes in daily weight, accurate data recording in an individualized home monitoring log book, and comprehensive review of predetermined physiologic parameters for which a breach requires immediate reporting. Furthermore, parents are provided contact information for the interstage cardiac care team to report concerns.

HMP

As originally described, the HMP is dependent on parental involvement, and includes discharge to home with a pulse oximeter (rental charge, \$350-\$580 per month) and infant scale sensitive within 10 g (rental charge, \$60-\$190 per month). Private insurance or Medicaid almost universally covered oximeter expenses, whereas scales were reimbursed

approximately 90% of the time. In the event scale coverage was not approved, scales obtained via donation were loaned to families. A parent or guardian recorded daily observations of SpO₂ and weight change. Through consensus of institutional cardiologists, cardiac intensivists, surgeons, and advance practice nurses, initial monitoring criteria necessitating notification to the interstage cardiac care team member included SpO₂ <75%, any weight loss of 30 g, or failure to gain 20 g over 3 days. Modifications to monitoring and subsequent notification were made based on quality improvement measures, and feedback solicited from families and providers. Specific modifications to notification criteria, including SpO₂ > 90 or intake < 100 cc/kg/d (June 2005) were implemented after an interstage death in 2005. Furthermore, the HMP expanded to include weekly interstage follow-up telephone contact (May 2004) between parents and the HMP team as well as development of a multidisciplinary cardiac specialty clinic for interstage infants (November 2005). At our institution, care of postoperative S1P infants through the interstage period is provided by a core group of cardiac intensivists, pediatric cardiologists, and cardiac nurse practitioners with daily management of HMP infants incorporated into the existing roles of these team members. No staff members have been added or exclusively designated to support the HMP.

Interstage events were defined as breach of the aforementioned arterial saturation or weight change criteria. Breach of predetermined parameters are immediately triaged by a member of the interstage cardiac care team who is available 24 h/d, 7 d/wk. As part of the triage process, physiologic trends, including the daily heart rate, are reviewed. The circumstances and severity of the interstage event prompt either follow-up telephone call within 24 hours, outpatient clinic or emergency room evaluation, and/or hospital admission. Parent-identified concerns beyond breach of saturation and weight criteria were addressed but not classified as events due to infrequent occurrence.

Data Collection and Analysis

Prospective perioperative and interstage databases were queried for the following variables: race; gender; birth weight; socioeconomic status; additional cardiac or extracardiac diagnosis; anatomic subtype; presence of aortic atresia; diameter of ascending aorta; age and shunt type at S1P; operative time; need for extracorporeal membrane oxygenator (ECMO) support; hospital length of stay; nutrition support, including need for enteral feeding tube; interstage breach of criteria events with interventions; and age and weight at S2P. Socioeconomic status was defined by the median income at residing ZIP code during the interstage period. Anatomic subtype was dichotomized as "HLHS" and "other" based on criteria established from the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease.¹⁹ Patients were considered high-risk at S1P if gestational age ≤ 35 weeks, birth weight < 2.5 kg, or additional cardiac or extracardiac diagnosis were present.

Data Analysis

Patient variables are summarized as count and percent, mean ± standard deviation, or median and range as appropriate. A bivariate analysis was performed comparing those with interstage events and those who did not breach criteria, and a second model compared those who remained as inpatients during the interstage period with those who were discharged home. A Student *t* test was used to test the difference between groups for continuous variables and Fisher exact test for categorical variables. Descriptions of interstage interventions are provided. Kaplan-Meier actuarial survival analysis with log-rank comparison between groups was used to compare overall survival. A significance level of *P* ≤ .05 was used throughout. Analysis was performed with IBM-SPSS version 20 (Armonk, NY).

RESULTS

From October 2000 to October 2010, 203 patients underwent S1P with 95% (192 out of 203) survival to hospital

FIGURE 1. Consecutive patients who underwent stage 1 palliation from October 2000 to October 2010. Transplant-free stage 1 palliation survivors discharged before stage 2 palliation participated in home monitoring with 98% interstage survival.

discharge or subsequent palliation: 157 (82%) S1P survivors were discharged from the hospital after S1P, 6 (3%) remained hospitalized until orthotopic heart transplant after S1P, and 29 (15%) remained as inpatients until S2P (Figure 1). All 157 patients discharged after S1P participated in the HMP and none were lost to follow-up. Five percent (8 out of 157) were non-English speaking. Health care coverage for HMP participants was equally distributed between private insurance and Medicaid. The modified Blalock-Taussig shunt (mBTs) was more commonly placed at S1P (100 out of 157; 64%) than the right ventricle to pulmonary artery shunt. Of the discharged cohort, 35 (22%) were considered high-risk patients.

Interstage Survival

Interstage survival was 98% (154 out of 157). Each interstage death occurred in standard-risk patients with mBTs, none of whom were discharged with enteral tube feeding. Two patients died suddenly without symptoms:

1 with greater than moderate atrioventricular valve insufficiency at age 30 days and 1 with acute shunt thrombosis diagnosed on postmortem evaluation at age 45 days. The third patient died after 12 hours of diminished oral intake and increased $\text{SpO}_2 > 90\%$ at age 28 days and a postmortem exam consistent with culture positive sepsis. This latter patient led to modification of criteria for notification to include $\text{SpO}_2 > 90\%$ and enteral intake $< 100 \text{ cc/kg/d}$. No patient since this incident presented with shock or respiratory failure during the interstage period.

Home Monitored Events

There were 156 home monitored events in 93 out of 157 patients (59%) (Table 1). Similar rates of events occurred in patients with the mBTs versus right ventricle to pulmonary artery shunt. Additionally, event rates were not statistically different between standard-risk and high-risk patients. In non-Hispanic white children, 62% (80 out of 129) had interstage events and in children of other racial and ethnic backgrounds 56% (15 out of 27) had events; this was not

TABLE 1. Patient and Norwood stage 1 palliation (S1P) perioperative characteristics in home monitoring program patients without and with events

	No event (n = 64)	Home event (n = 93)	P value
Birth weight (kg)	3.3 ± 0.6	3.2 ± 0.6	.5
Gestational age (wk)	38.3 ± 1.5	38.2 ± 1.2	.7
Male gender	40 (66)	56 (58)	.2
Race			.5
Non-Hispanic white	49 (80)	80 (83)	
Non-Hispanic black	8 (13)	2 (2)	
Hispanic	4 (7)	7 (7)	
Asian/American Indian	0 (0)	6 (6)	
Income at residing ZIP code (median \$ [IQR])	\$52,000 (\$45,000-\$65,000)	\$52,000 (\$41,000-\$70,000)	.4
Risk status at S1P			.2
High-risk	11 (18)	25 (26)	
Standard-risk	50 (82)	71 (74)	
HLHS	48 (78)	80 (83)	.25
Aortic atresia	30 (82)	53 (55)	.2
Ascending aorta diameter (mm)	3.7 ± 1.7	3.5 ± 1.7	.4
Age at S1P (d)	8.2 ± 8.7	7.5 ± 4.9	.5
Total support time (min)	188 ± 52	182 ± 42	.5
Cardiopulmonary bypass (min)	181 ± 46	170 ± 42	.14
Circulatory arrest (min)	10 ± 5	12 ± 6	.17
Shunt type			.4
Modified Blalock-Taussig shunt	40 (66)	60 (62)	
Right ventricle-pulmonary artery shunt	21 (34)	36 (38)	
ECMO support	3 (5)	5 (5)	.6
S1P hospital length of stay (d)	38 ± 18	40 ± 20	.4
Feeding method at S1P discharge			.08
Orally fed	38 (62)	63 (66)	
Exclusive enteral tube fed	1 (2)	9 (9)	
Partial enteral tube fed	22 (36)	24 (25)	

Values are given as mean ± standard deviation or n (%), unless otherwise noted. *IQR*, Interquartile range; *S1P*, stage 1 palliation; *HLHS*, hypoplastic left heart syndrome; *ECMO*, extracorporeal membrane oxygenator.

a significant difference ($P = .53$). Socioeconomic status based on median income at residing ZIP code was similar between those patients with and without events (median, \$52,000; interquartile range, \$45,000-\$65,000 vs median, \$52,000; interquartile range, \$41,000-\$70,000; $P = .4$). Nearly one-third of all HMP patients (50 out of 157) had 1 event, 28 (18%) had 2 events, 10 (6%) had 3 events, and 5 (3%) had > 3 events. Of 93 patients who had either single or multiple events, 69 (74%) experienced at least 1 SpO₂ event, 38 (41%) experienced at least 1 weight event, and 8 (9%) had concurrent SpO₂ and weight events. Nearly all patients with events (90 out of 93) were readmitted at least once during the interstage period with a median readmission rate of 1 (range, 1-5). In patients with events that led to hospital readmission, median duration from S1P hospital discharge to readmission was 32 days (range, 2-144 days). Patient management following reported events is shown in Figure 2. The majority of readmissions triggered by interstage events led to catheterization or supportive medical therapy. Specific medical therapies included rehydration, antibiotics, blood transfusion, gastrointestinal tube placement, and/or respiratory

management. Early S2P was performed in 36 patients who were readmitted.

Pulse oximetry <75% was the most common reason for breach of predetermined home monitored criteria. The etiology of progressive hypoxia was unclear in one-third of patients; however, almost 30% of patients with SpO₂ events were found to have a respiratory illness (Table 2). Of the 22 patients with progressive hypoxia secondary to anatomic lesions, 17 had shunt or brachiocephalic narrowing, 4 had moderate or greater atrioventricular valve insufficiency, and 1 had a restrictive atrial septum defect.

In the subset of patients with breach of weight criteria, inadequate enteral intake < 100 cc/kg/d was the most frequent cause, accounting for 45% of events (Table 2). Increasing caloric density or enteral tube feeding volume in patients with gastrostomy tubes were the most common interventions for failure to gain sufficient weight. Approximately one-third (54 out of 157) of the home monitored cohort had surgical gastrostomy tubes placed before discharge following S1P, 3 were readmitted for placement of a gastrostomy tube and were discharged back to home, and 6 remained hospitalized after readmission

FIGURE 2. Interstage management for home monitored patients with outpatient events. Events are defined as oxygen saturation <75%, any weight loss of 30 g, or failure to gain 20 g in 3 days. Additional event criteria added in 2005 are oxygen saturation >90% and enteral intake < 100 cc/kg/d.

for nasogastric feeds due to inconsistent oral intake and close proximity to planned S2P. Average interstage growth velocity for the HMP cohort from S1P hospital discharge to S2P was normal for infant growth at 26 g/d.

S2P

Age at S2P was younger in patients with home monitored events compared with patients without events (112 ± 29 vs 135 ± 37 days; $P < .001$). Patients with events weighed less at S2P compared with patients without events (5.51 ± 0.90 vs 5.82 ± 0.78 kg; $P = .03$). However, weight-for-age z score was similar between patients with and without events (-0.57 ± 1.3 vs -0.38 ± 1.3 ; $P = .57$). Hospital length of stay after S2P was longer in patients with events (median, 12 days; range, 4-235 days) compared with those without events (median, 8 days; range, 3-105 days; $P = .0003$). Overall survival to hospital discharge after S2P for the home monitored cohort was 100%. One-, 5-, and 10-year survival by event status is shown in Figure 3.

TABLE 2. Etiologies associated with 156 home-detected events in 93 patients

	SpO ₂ events (n = 93)	Weight events (n = 55)	SpO ₂ and weight events (n = 8)
Anatomic lesion	22 (24)	0	3 (37.5)
Respiratory illness	26 (28)	3 (6)	0
Anemia	8 (9)	0	0
Inadequate intake	2 (2)	25 (45)	3 (37.5)
Emesis	5 (5)	14 (25)	0
Diarrhea	0	3 (6)	2 (25)
Nonspecific	30 (32)	10 (18)	0

Values are given as n (%). SpO₂, Oxygen saturation.

Interstage Inpatient Management

Fifteen percent (29 out of 186) of transplant-free S1P survivors remained hospitalized until S2P (Table 2); this cohort differed from those discharged during the interstage period in a number of variables, as shown in Table 3. Of these 29 inpatients, 13 (45%) had high-risk characteristics at S1P, 20 (69%) received an mBTs at S1P, and 9 (31%) required ECMO. The inpatient cohort had longer intraoperative support time with a trend toward having a diagnosis other than HLHS and older age at S1P. Reasons for inpatient management throughout the interstage period included labile systemic perfusion in 17 patients (7 with atrioventricular valve insufficiency, 1 with recurrent arch obstruction, 4 with arrhythmias, and 5 with nonspecific findings); labile SpO₂ in 4 patients; and extracardiac organ dysfunction in 8 patients. For the interstage inpatients, age at S2P was 107 ± 42 days and weight at S2P was 4.7 ± 1.5 kg. Survival to hospital discharge after S2P was 83% (24 out of 29). Intermediate survival beyond S2P is shown in Figure 3.

DISCUSSION

The HMP after Norwood S1P for HLHS and its variants began in response to interstage mortality within our program despite marked improvement in operative survival.¹⁸ The fundamental design of the HMP is a simple, inexpensive, and effective method of empowering and supporting parents in the home environment to prevent interstage death through the early detection of subtle physiologic changes that may precede overt clinical deterioration in an infant's condition.^{6,7,18} The unintended benefits were earlier recognition of inadequate growth and the elimination of growth failure that is commonly reported in infants with shunt-dependent heart disease.^{14,18,20,21}

FIGURE 3. Actuarial survivals for home monitored patients with events, home monitored patients without events, and patients who remained hospitalized to stage 2 palliation (S2P). Survivals beyond S2P are similar between home monitored patients with and without interstage events ($P = .3$). Survivals beyond S2P are lower in interstage inpatients than those discharged to home with and without events ($P = .001$ for both).

After a decade of interstage home monitoring, we observed 98% interstage survival in this cohort with no deaths in discharged patients since 2005. Similarly, since our last interstage death at our institution in 2005, no patient presented in severe shock or with respiratory failure. However more than half of discharged patients had timely detection of interstage desaturation or concerning weight changes prompting evaluation. The majority of home monitoring events led to admission and, commonly, these infants had diagnoses associated with interstage death in previous reports.

In the multicenter SVR trial, risk factors for interstage death included gestational age < 37 weeks, Hispanic ethnicity, poverty level, diagnosis of aortic atresia/mitral atresia, an increased number of postoperative complications, and a mBTs in the presence of less than moderate atrioventricular valve insufficiency.⁶ In our cohort, several of these variables associated with death were evaluated and were not found to be risk factors for interstage events. Specifically, prematurity, HLHS diagnoses, aortic atresia, ECMO, S1P hospital length of stay, and shunt type were similar between discharged

TABLE 3. Patient and stage 1 palliation (S1P) perioperative characteristics in home monitoring program (HMP) patients and interstage inpatients

	Home monitored (n = 157)	Inpatient to S2P (n = 29)	P value
Birth weight (kg)	3.2 ± 0.6	3.1 ± 0.7	.4
Gestational age (wk)	38.3 ± 1.3	38.0 ± 1.8	.4
Male gender	96 (61)	15 (51)	.2
High risk status	36 (23)	13 (45)	.015
HLHS	128 (82)	19 (66)	.08
Aortic atresia	83 (53)	10 (35)	.07
Ascending aorta diameter (mm)	3.6 ± 1.7	3.9 ± 2.0	.4
Age at S1P (d)	7.8 ± 6.6	10.6 ± 10.9	.06
Total support time (min)	185 ± 46	208 ± 81	.04
Cardiopulmonary bypass (min)	175 ± 44	202 ± 67	.007
Circulatory arrest (min)	11 ± 6	13 ± 10	.08
Modified Blalock-Taussig shunt	100 (64)	20 (69)	.4
ECMO support	8 (5)	7 (24)	.002

Values are given as mean ± standard deviation or n (%). S2P, Norwood stage 2 palliation; HLHS, hypoplastic left heart syndrome; ECMO, extracorporeal membrane oxygenator.

patients with and without events. Socioeconomic status and race were not associated with the occurrence of interstage events. Atrioventricular valve insufficiency, and thus its relationship with other previously reported risk factors such as shunt type, was not evaluated in this study. Such an analysis would likely be biased by our programmatic approach to managing the most symptomatic patients, including those with greater than moderate atrioventricular valve insufficiency, as inpatients throughout the interstage period. Nearly a quarter of home monitored patients in our cohort had high-risk characteristics. The lack of differences in our event rates between high-risk and standard-risk patients may reflect use of a structured HMP regardless of risk status, unlike interstage management in the SVR trial.

Method of feeding, specifically exclusive tube feeding and nasogastric versus gastric feeding, has been identified as an interstage risk factor through univariate analysis alone in the SVR trial.⁶ Multicenter adoption of our HMP facilitated by the National Pediatric Cardiology-Quality Improvement Collaborative led to identification of nasoenteral tube feeding as an associated risk factor for interstage death.¹⁴ In our series, the number of patients who were orally fed was similar between groups; however, there was a trend toward more interstage events in patients who were fed via a surgical gastrostomy tube. This observation likely reflects early practice at a time when the interstage team did not consistently manage feeds. No patient was discharged with a nasogastric tube per institutional practice.

In this series we were unable to identify a subset of this vulnerable cohort most likely to have interstage events and thus benefit from a HMP of SpO₂ and weight change before admission for S2P. Therefore, these results highlight the ongoing benefit of interstage home monitoring for all patients discharged from the hospital after S1P. The absolute benefit of an HMP is unknown given the absence of a control group during this era. Nevertheless, the strength of these data resulted in expansion of our program in 2005 to every infant with shunt-dependent circulation regardless of ventricular morphology, similar to a structure reported by Dobrolet and colleagues.²²

Similar to the SVR trial interstage report, a subset of patients deemed to be at high risk for interstage mortality remained as inpatients until S2P. Specifically the interstage inpatients more often had high-risk features (prematurity, low birth weight, extracardiac disease, or additional cardiac diagnoses) and demonstrated physiologic instability with regular infant care. Intraoperative support times were longer and the need for postoperative ECMO support greater in the interstage inpatients. Whereas aortic atresia independent of mitral valve pathology was associated with interstage death in the SVR trial and other single-center reports,⁶ we observed that interstage inpatients had a trend in a diagnosis

other than HLHS and were less likely to have aortic atresia. The increased vulnerability in the patients who remained as inpatients until S2P is further illustrated in their reduced survival results beyond the cavopulmonary shunt connection compared with the patients deemed safe for discharge after S1P (Figure 3).

Desired thresholds for SpO₂ and weight change were determined at inception of the HMP and derived through consensus opinion from cardiologists, cardiac intensivists, cardiothoracic surgeons, and advanced practice nurses. Based on initial experiences, patient outcomes, and parental input, the scope of the program was broadened for detection of circulatory insufficiency and failure to thrive. Concomitantly, several program modifications were instituted to optimize effectiveness of this in-home surveillance strategy. Specifically, nutrition goals, standardized equipment, structured data-recording, educational tools, and room-in care by parents was implemented before discharge. Weekly telephone contact was made by an interstage nurse to provide parental support, assess nutrition and growth, and assess pulse oximetry trends. A routine interstage specialty clinic staffed by a dedicated cardiac care team of physicians, nurse practitioners, dietitians, and feeding experts was established to provide in-person assessments of these complex infants by a consistent care team. At interstage clinic visits, routine imaging and laboratory data are not routinely performed but rather obtained as needed based on clinical status. Patient progress determines timing for pre-S2P echocardiogram and cardiac catheterization.

A growing body of evidence supports the application of the medical home principle to high-risk pediatric populations. The key principle embodied in the medical home concept is care coordination to aid in the management of medically complex children. The HMP was provided for all patients at S1P hospital discharge, and hence the absence of a contemporaneous control group and interstage inpatient management for a select subset limits our ability to assess causation in the relationship of home monitoring practices and improved interstage outcomes. However we have learned from previous reports that using medical home principles, specialized ambulatory care, and care coordination in at-risk patients is associated with streamlining care for children and families; optimizing the use of medical resources; expanding the competence of involved providers; increasing parent satisfaction; and, most importantly, improving patient outcomes.²³⁻²⁵ Specific improvements to this cohort include better somatic growth, near elimination of patients who present to the hospital in extremis, and improved interstage survival.

CONCLUSIONS

Home monitoring of SpO₂, weight change, and enteral intake as part of an interstage surveillance program that

also includes weekly telephone contact and multidisciplinary clinic visits is effective in identifying at-risk physiologic changes in infants following SIP. After a decade of experience, this program continues to be associated with excellent interstage survival similar to the initial report of interstage home monitoring during 2003.¹⁸ The added benefit generated from the HMP is a greater focus on optimizing nutrition during the interstage period and normalized weight gain in a historically undernourished infant group. The aforementioned interstage program effectively addresses the comprehensive care coordination needs of this medically complex patient population consistent with US Department of Health and Human Services Healthy People 2010 goals and objectives, which state that “all children with special health care needs will receive regular ongoing comprehensive care within a medical home.”²⁶

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