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Review

# Interstage monitoring: Yes it makes a difference!

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ABSTRACT

Hypoplastic left heart syndrome is associated with a substantial interstage mortality between the Stage I neonatal Norwood operation and the Stage II bidirectional cavopulmonary anastomosis. One of the many endeavors to reduce interstage mortality has been the development of interstage home monitoring programs. We review the efficacy of interstage monitoring programs in general and in particularly at our center and conclude that interstage monitoring does make a difference. All programs, however, are not the same. Further research is needed to ascertain which facets of interstage monitoring truly impact outcomes and restructure monitoring programs as such.

Hypoplastic left heart syndrome remains one of the most challenging lesions to manage in all of congenital heart disease, with a significant out-of-hospital, "interstage" mortality between the neonatal operation (Stage I, Norwood) and the second stage bidirectional cavopulmonary anastomosis at 4 to 6 months of age. The multicenter Single Ventricle Reconstruction trial (2005–2008) reported a 69% survival rate at 12 months [1] for infants undergoing staged palliation for hypoplastic left heart syndrome with a 12% mortality [2] rate during the interstage period.

One of many endeavors proposed to address this interstage mortality has been the development of at-home interstage monitoring programs. In 2003 Ghanayem, et al. described the first interstage monitoring program at the Children's Hospital of Wisconsin with a decrease in interstage mortality from 15.8% pre-program to 0% 14 months into program implementation [3]. The main interventions of the program included sending infants home with a weighing scale and pulse oximeter machine. Parents were instructed to maintain 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%, an acute weight loss of more than 30 g in 24 h, or failure to gain at least 20 g during a 3-day period.

Similar programs subsequently developed across the country. Currently most of the 65 US centers in the National Pediatric Cardiology Quality Improvement Collaborative, a national initiative focused on reducing mortality during the interstage period, have ongoing interstage programs. The purpose of this paper is to review the efficacy of interstage monitoring programs in general and in particularly at our center with one of the largest populations of hypoplastic left heart syndrome infants in the country.

In 2012, Feinstein led a white paper publication from 15 US centers as well as The Hospital for Sick Children, Toronto, CA and the Royal Brompton, London, UK [4]. They concluded that the current best practice for interstage care is heightened surveillance of this at-risk population through participation in a home monitoring program. They hypothesized that early, at-home detection of variation in the measured parameters may indicate the development of serious anatomic lesions or an intercurrent illness and allow for life-saving interventions.

Since the 2012 white paper, seven single center and one multicenter reports have been published describing results of interstage monitoring programs in the US and Europe (Table 1) [5–12]. Six of eight report significant reduction in mortality with post-initiation of interstage program mortality rates ranging from 0 to 8% [5–7,9,11,12]. On the contrary, Oster in 2015 analyzed the National Pediatric Cardiology Quality Improvement Collaborative data from 2008 to 2012 and found no association of saturation and weight monitoring with interstage mortality [8]. Likewise, in a single center study from the Children's National Health System there was no change in mortality but statistical improvement in weight at Stage II and complications post Stage II were found [10].

The most recent publication on this topic is from our center at the Children's Hospital of Philadelphia [12]. Our interstage program was initiated in December 2010. In our retrospective cohort study historical controls (2007–2010) were compared to interstage monitoring program patients (2011–2015) with a decrease in overall interstage mortality for

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**Table 1**
Recent reports on interstage monitoring programs.

| Publication date | Author | Institution | Study dates pre-ISM to post-ISM | Interstage mortality pre-ISM to post-ISM |
|---|---|---|---|---|
| 2012 | Hansen [5] | Kiel, Germany | 1996–2004 to 2005–2009 | 12.4% to 2.2% |
| 2014 | Rudd [6] | Children's Hospital of Wisconsin | 1996–2000 to 2000–2010 | 15.8% [3] to 2% |
| 2014 | Siehr [7] | Stanford | 2005–2010 | 0% interstage mortality |
| 2016 | Castellanos [9] | Children's Hospital of Los Angeles | 2007–2009 to 2010–2012 | 18.9% to 3.8% |
| 2017 | Texter [11] | Nationwide Children's Hospital | 2010–2013 to 2014–2016 | 17% 8% to (0% in 2016) |
| 2019 | Gardner [12] | Children's Hospital of Philadelphia | 2007–2010 to 2011–2015 | 14% to 4.8% (0% since April 2016) |
| 2015 | Oster [8] | NPC-QIC (multicenter) | 2008–2012 | No association of saturation or weight monitoring with interstage mortality |
| 2016 | Harahsheh [10] | Children's National Health System | 2005–2008 to 2009–2014 | No improvement in interstage mortality but improvement in weight at Stage II and complications post Stage II |

ISM (interstage monitoring program).
Reports from interstage monitoring programs from 2012 to 2019 are listed above. The first 6 reports show improvement in mortality after interstage monitoring programs were initiated at the respective center.

all single ventricle infants from 13% to 5.4% ($p = 0.02$) and for hypoplastic heart syndrome alone 14% to 4.8% ($p = 0.03$). We have sustained these excellent results with a 0% interstage mortality for hypoplastic left heart syndrome infants since April 2016.

In addition to discharge with a pulse oximeter and weighting scale, the components of our program include:

1. Standardization of the neonatal discharge criteria and process
2. Standardization of parental education prior to discharge including a list of “red-flags” to trigger immediate contact of the interstage program by the parents
3. Daily home monitoring by parents including daily oxygen saturation measurements, daily weight measurements, and feeding log
4. At least weekly home nursing visits documenting oxygen saturation, weight, mode of feeding, formula and daily amount
5. At least weekly phone calls to parents by a dedicated interstage monitoring program nurse practitioner with necessary adjustment in feeding plans and referral to pediatrician, primary cardiologist or emergency department if needed
6. Weekly assessment of individual feeding plans by a program registered dietitian
7. Bi-weekly pediatrician visits
8. Bi-weekly (after Norwood) or monthly (other shunt dependent patients) primary cardiology visit with focused echocardiograms either at The Children's Hospital of Cardiology or locally
9. Weekly review of each patient by the dedicated interstage monitoring program team
10. Scheduling of standard Stage II operation at 4–5 months of age.

Even though we have a dedicated interstage monitoring team that includes 5 program cardiologist and a dedicated nurse practitioner, we do not have a single ventricle clinic. Communication and co-management are ongoing throughout the interstage period among the interstage team members, particularly the dedicated nurse practitioner, and the primary cardiologist either at the Children's Hospital of Philadelphia or locally.

The most recent studies, including our own, support the conclusions of the 2012 white paper [4] that interstage monitoring does make a difference and should be recommended. It must be noted, however, that all interstage monitoring programs are not the same. Monitoring of daily weights and pulse oximeter readings is not the entire picture. Weight and saturation measurements are necessary but not sufficient to monitor ***and manage*** these fragile infants. From the description of our program above, one can see that it is much more than tracking daily weights and saturations. In our experience it is the close professional relationship that develops between the program nurse practitioner (as the representative of the interstage team) with the family on the one hand, and with the primary cardiologist on the other hand that is paramount to early identification of potential life-threatening events. It is the ***management*** as well as the monitoring of these fragile infants that is key.

In addition, learning has occurred over time in both individual centers as well as in the National Pediatric Quality Improvement Collaborative regarding interstage monitoring. This may well explain the lack of positive effect of interstage monitoring in Oster's analysis of National Pediatric Quality Improvement Collaborative data from 2008 to 2012, early in the collaborative experience. Much intra and inter-institutional learning has occurred since then. A current analysis may well give a different picture, especially since a center line shift occurred in interstage mortality in the collaborative in August 2013 from 9.5% to 5.1% and has been sustained [13].

What is the future of interstage monitoring? It is here to stay with potential for substantial growth. Many centers have broadened interstage monitoring to include monitoring of other fragile infants with complex congenital heart disease such as complex Tetralogy of Fallot with pulmonary atresia. In addition, some centers as well as ours are embarking on telemedicine [10,14] in this patient population as well as predictive analytics (artificial intelligence) to predict critical events both in-hospital [15] and at home. Finally, home monitoring is primed to expand beyond the interstage period with current endeavors of the National Pediatric Cardiology Quality Improvement Collaborative to monitor patients from fetal life through Fontan palliation (https://npcqic.org).

In conclusion, interstage monitoring does make a difference. All programs, however, are not the same. We as a pediatric cardiology community needed to ascertain which facets of monitoring truly impact outcomes and restructure our monitoring programs as such.

## Declaration of Competing Interest

Therese Giglia: none
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Monique Gardner: none
Shobha Natarajan: none
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Jonathan Rome: none

## References


[1] Ohye RG, Sleeper LA, Mahony L, et al. Comparison of shunt types in the Norwood procedure for single-ventricle lesions. N Engl J Med 2010;362:1980–92.

[2] Ghanayem NS, Allen KR, Tabbutt S, et al. Interstage mortality after the Norwood procedure: results of the multicenter single ventricle reconstruction trial. J Thorac Cardiovasc Surg 2012;144:896–906.

[3] Ghanayem NS, Hoffman GM, Mussatto KA, et al. Home surveillance program prevents interstage mortality after the Norwood procedure. J Thorac Cardiovasc Surg 2003;126:1367–77.

[4] Feinstein JA, Benson DW, Dubin AM, et al. Hypoplastic left heart syndrome: current considerations and expectations. J Am Coll Cardiol 2012;59:S1–42.

[5] Hansen JH, Furck AK, Petko C, et al. Use of surveillance criteria reduces interstage mortality after the Norwood operation for hypoplastic left heart syndrome. Eur J Cardiothorac Surg 2012;41:1013–8.

[6] Rudd NA, Frommelt MA, Tweddell JS, et al. Improving interstage survival after Norwood operation: outcomes from 10 years of home monitoring. J Thorac Cardiovasc Surg 2014;148:1540–7.

[7] Siehr SL, Norris JK, Bushnell JA, et al. Home monitoring program reduces interstage mortality after the modified Norwood procedure. J Thorac Cardiovasc Surg 2014;147:718–23 e1.

[8] Oster ME, Ehrlich A, King E, et al. Association of Interstage Home Monitoring with Mortality, readmissions, and weight gain: a multicenter study from the National Pediatric Cardiology Quality Improvement Collaborative. Circulation 2015;132:502–8.

[9] Castellanos DA, Herrington C, Adler S, Haas K, Ram Kumar S, Kung GC. Home monitoring program reduces mortality in high-risk sociodemographic single-ventricle patients. Pediatr Cardiol 2016;37:1575–80.

[10] Harahsheh AS, Hom LA, Clauss SB, et al. The impact of a designated cardiology team involving telemedicine home monitoring on the Care of Children with single-ventricle physiology after Norwood palliation. Pediatr Cardiol 2016;37:899–912.

[11] Texter K, Davis JAM, Phelps C, et al. Building a comprehensive team for the longitudinal care of single ventricle heart defects: building blocks and initial results. Congenit Heart Dis 2017;12:403–10.

[12] Gardner MM, Mercer-Rosa L, Faerber J, et al. Association of a Home Monitoring Program with Interstage and Stage 2 outcomes. J Am Heart Assoc 2019;8:e010783.

[13] Anderson JB, Beekman 3rd RH, Kugler JD, et al. Improvement in Interstage survival in a National Pediatric Cardiology Learning Network. Circ Cardiovasc Qual Outcomes 2015;8:428–36.

[14] Fleck DA, Marino BS, Costello JM, et al. The REACH protocol: an innovative strategy for home management of infants with complex CHD. Cardiol Young 2018;28:961–7.

[15] Ruiz VM, Saenz L, Lopez-Magallon A, et al. Early prediction of critical events for infants with single-ventricle physiology in critical care using routinely collected data. J Thorac Cardiovasc Surg 2019;158(1):234–243.e3. https://doi.org/10.1016/j.jtcvs.2019.01.130. Feb 21.PMID: 30948317.