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Review

Gaps in pediatric cardiac care: Opportunities to make a difference

Matthew D. Files[a,*], Lori Erickson[b], Kathleen Reid[c], Lisa Roten[d], Adrea Leach[e], Jo Ann Davis[f], Dongngan Truong[g], Girish Shirali[b]

[a] Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105, United States of America
[b] The Ward Family Heart Center-Children's Mercy Kansas City, 2401 Gillham Road, Kansas City, MO 64108, United States of America
[c] Children's National Health System, 111 Michigan Avenue NW, Washington, DC 20010, United States of America
[d] Cook Children's Medical Center, 800 7th Avenue, Ft Worth, TX 76104, United States of America
[e] WVU Medicine Children's, 1 Medical Center Drive, Morgantown, WV 26506, United States of America
[f] Nationwide Children's Hospital, 700 Children's Dr., Columbus, OH 43205-2664, United States of America
[g] Primary Childrens Hospital, 81 N Mario Capecchi Dr, Salt Lake City, UT 84113, United States of America

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ABSTRACT

Dramatic surgical and medical advances in congenital heart disease over the past 50 years have led to progressive increases in survival for children born with heart defects, with the vast majority now expected to survive into adulthood. Children with the most severe spectrum of congenital heart disease, those with functionally univentricular hearts, continue to face a significant burden of morbidity and mortality, particularly in the first 6 months of life. This review summarizes a strategy to address the gaps in care for these children by examining the 7-year experience with a novel, tablet and cloud-based home monitoring program called CHAMP. We explore the gaps in care that we sought to address, as well as unexpected opportunities that we discovered during this experience.

1. Introduction

Over half a century has passed since Dr. Rashkind devised the balloon atrial septostomy, which he described, rightly, as the world's fastest operation. The prognosis for congenital heart defects has truly been transformed over these 50 years. Defects that were considered inoperable are now routinely managed surgically. One good way to quantify the improvement in prognosis is to examine long-term, prospective, population studies. Erikssen et al. [1] reported on the follow-up of all 7038 children who underwent cardiac surgery in Oslo, Norway, over the course of 40 years between 1971 and 2011. The study reports on all-cause mortality until 2012. The improvement in survival has been dramatic, with 1-year survival, which used to be around 71% in the mid-1970s, rising to over 99% by 2011. Studies such as this actually underestimate improvements, because they do not account for the patients for whom surgery was not even an option in the 1970s or 1980s. Overall, thanks to advances in knowledge, skill, teamwork and technology, our patients and families today can look forward to a much better prognosis than ever before. But gaps remain, representing opportunities to shift the paradigm and set the stage for improvements that may be coming. The management of interstage infants with a functionally univentricular heart is an example of one such area. We have previously described a novel tablet and cloud-based home monitoring program for interstage infants (Cardiac High Acuity Monitoring Program, CHAMP)® [2,3]. The current paper is a summary of the evolution of our center's 7-year experience with CHAMP, including the multi-center rollout of the program.

2. Background

Infants born with univentricular heart disease represent the most challenging spectrum of congenital heart disease able to be palliated. This population continues to experience high rates of morbidity and mortality, particularly in early childhood [4–7]. These children require a staged surgical palliation with 3 operations, the first of which is performed in the first days after birth. The second operation (superior cavopulmonary anastomosis, the Glenn operation) is typically performed between 4 and 6 months, and the final stage (total cavopulmonary anastomosis, the Fontan operation) between 2 and 5 years. The period between discharge from the first operation and the second, termed the “interstage period,” represents a period of significant vulnerability with high rates of hospital readmission and mortality rates between 5 and 20% [8,9]. In 2000, Ghanayem et al. implemented a program of home monitoring during this interstage period [8]. Parents

* Corresponding author.
E-mail address: matthew.files@seattlechildrens.org (M.D. Files).

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recorded daily weights and oximetry into a daily log which was reviewed at each clinic visit, and parents were instructed to call the care team with concerns or if certain threshold values were breached. This program was associated with a drastic reduction in interstage mortality. Many centers around the country have adopted similar home monitoring programs to confront interstage mortality. Unfortunately, interstage morbidity and mortality remain disappointingly high today.

3. NPCQIC and expansion of home monitoring programs

In recognition of the problem of interstage mortality following the Norwood procedure, the Joint Council on Congenital Heart Disease convened the National Pediatric Cardiology Quality Improvement Collaborative (NPCQIC) in 2008 to specifically work to improve interstage survival [10]. The collaborative encouraged home monitoring as described above while also promoting centers to develop standard protocols, dedicated care teams, with many implementing focused high-risk outpatient clinics [11]. Despite robust participation from care teams and parents in the NPCQIC collaborative, recent publications from 2018 suggest high ongoing morbidity (50% reintervention rate [12]) and 5–10% interstage mortality [11,13,14] within this patient population. Clearly, there is still room for improvement for this vulnerable group.

4. Limitations of current home monitoring

One of the limitations of paper-based approaches is that they place the burden of data collection, recording and interpretation on the parents or caregivers. The family is also responsible for triage of perceived problems and contacting the care team if concerned. This is in addition to the overwhelming stresses that come with having a newborn infant, and managing multiple medications, complex feeding schedules and equipment, and all of the other burdens that come with having a medically complex infant at home [15]. There is a real risk of delay in care and loss of data. Overall, the paper-based approach represents a reactive model of home care, rather than a proactive process.

5. Design of CHAMP

Our main goals in restructuring a home monitoring platform for this population were to reduce the burden of data collection, transmission, and analysis by the family, and to reduce delays by having that data immediately available to the care team. We also wanted to move the triage process from the family to the care team by sending instant alerts, and eventually, to use predictive analytics to determine which patients were at increasing risk of decompensation. The design for CHAMP has been previously described [3], but briefly we built an informatics infrastructure from the ground up at Children's Mercy Kansas City (CMKC). The caregiver at home records home monitoring data and videos in a simple, tablet-based application which is transmitted to a secure, HIPAA-compliant cloud-based database. This database then interfaces with real-time analytics, and transmits data to the care team via instant alerts to the pagers or cell phone of the care team, who can view data through a secure web portal. Data are also transmitted to the electronic medical record. We also included the ability to record a 15 s, high-definition video, and asked families to record daily videos of their child awake and undressed, so that the care team could observe rate and work of breathing, interactivity, and overall clinical state on a serial basis.

The web-based portal for the care team was built to visually display home monitoring data graphically, (Fig. 1). Additionally, data from readmissions and red-flag alerts is overlaid with home monitoring data to visually recognize trends and clinically significant events. We felt this represented a significant improvement over the paper-based system (as well as the electronic medical record).

Fortunately, the funding for CHAMP has been provided by philanthropy from the Claire Giannini Foundation and CMKC, and as such, families receive this monitoring equipment (with cellular connection) at no cost. The user interface was designed with input from parents; it is simple and intuitive and has been translated into 9 languages at low health literacy reading levels for all languages. This is an important step to mitigate significant racial and socioeconomic inequality, which has long been associated with poor outcomes in chronic disease, and has recently shown to be an important predictor of transplant-free survival for Norwood patients [16].

6. CHAMP crossover study

We recently published a randomized crossover study comparing CHAMP to the traditional 3-ring binder method in 31 infants [2]. Following discharge after neonatal palliation, home monitoring consisted of a 3-ring binder for the first month after neonatal discharge. After the first month, they were randomized to either stay with 3-ring binder or change to the tablet-based CHAMP program, and as such the patients served as their own control. After the second month, those who had the 3-ring binder switched to CHAMP. After families had experienced each method for 1 month, they were given the choice between CHAMP and the 3-ring binder. There was no interstage mortality in these 31 infants. During CHAMP monitoring, ICU and hospital readmissions were shorter, as were delays between the recording of a clinical change to communication to the team and admission to the hospital. Growth failure, which is a strong surrogate for overall health and survival in infants with univentricular hearts [17], was decreased during CHAMP monitoring. Additionally, 13/23 (57%) unplanned readmissions during CHAMP monitoring were based on CHAMP data instead of caregiver concerns, which suggests that the care team was responding in a proactive manner. Finally, CHAMP was preferred by the majority of caregivers.

The crossover study demonstrates that a key benefit of CHAMP is reducing delays in care, which can be particularly devastating for these children. Prior work has classified these delays as a delay in recognizing a symptom, delays in deciding if a symptom suggests illness, and a delay in obtaining care after decision to seek care [18]. We feel the structure of CHAMP addresses each of these domains.

To date, 129 children have been monitored with CHAMP at CMKC with a 3.1% interstage mortality. Post-discharge transplant free survival has significantly improved for all univentricular patients ($p = 0.02$) as well as Norwood patients ($p = 0.02$) compared to historical controls from 2010 to the current time (unpublished data). There is no doubt that there are many other concurrent clinical changes in the surgical, ICU, and outpatient care that may well account for improved mortality in these infants (era effect).

7. National rollout

While the outcomes for CHAMP at CMKC have been promising, we wanted to address questions about whether these outcomes could be scaled to other centers. We also wanted to collate data from multiple sites in order to enhance the statistical power of the program. Starting with Seattle Children's Hospital in 2016 and expanding significantly over the past two years (Fig. 2), there are currently 8 centers and a total of 370 patients who have participated; 223 of these have completed their Glenn operation. Collectively there is a 2.4% interstage mortality rate (some of these infants are still interstage) which suggests that we have been able to scale to a national level and replicate our results in different centers.

Costs of the rollout to partner sites have been funded by continued philanthropic support by the Claire Giannini Foundation as well as by CMKC. Each center has had to modify their practice to successfully navigate their new real-time interactions with clinical data, videos, and instant alerts. This increased vigilance and continuous reporting may have required adjustments to the single ventricle team staffing. All

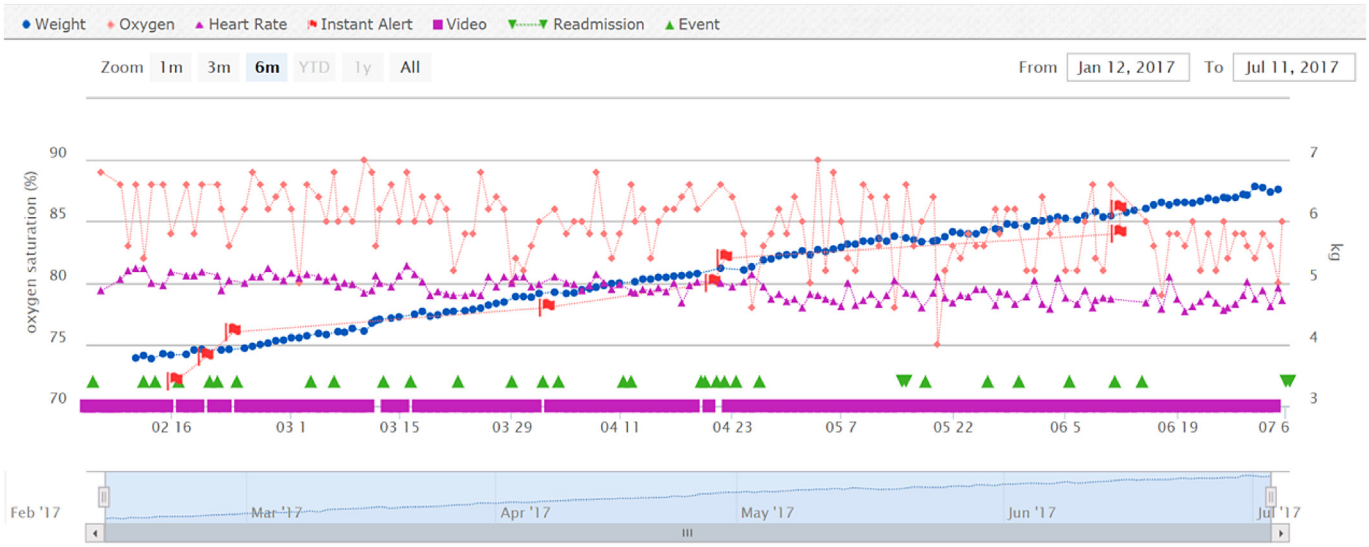

Fig. 1. Sample of clinical data as viewed through the CHAMP webportal. Clinical data is displayed over time to visualize trends as well as recognize important clinical events in relation to home monitoring parameters. Weight is recorded in blue circles, oxygen saturation in purple triangles, heart rate in pink squares, with clinic visits as green triangles and hospital readmissions as inverted triangles. The data-triggered red flag events are also visualized.

partner sites have started with a full day of on-site training at CMKC, consisting of hands-on review of CHAMP, the CHAMP Webportal, and clinical use. We continue to iteratively revise training manuals for families, caregivers, and information technology aspects. We provide direct communication for care teams at other sites through our front-line clinical nursing team, and Heart Center Informatics support (as needed) during the initiation of CHAMP applications at the partner sites.

We collect a robust amount of clinical data about patient characteristics, hospitalizations, operative details and clinic visits as part of the CHAMP database. Teams have needed to dedicate personnel and resources that are responsible for this data collection. Many participating centers already had – or have now developed – a dedicated interstage team of providers who care for these children. As such, it is challenging to separate the clinical effect of the CHAMP home monitoring from the additional infrastructure that comes with a dedicated interstage care team.

8. Video analysis

Shortly after our program launched, we asked parents to record daily videos which can be compared if there is a clinical change. The decision to add video was serendipitous (the tablet PC had video capability). The additive impact of videos to the overall assessment the child's clinical status has been an unexpected benefit. We have found that much can be learned about an infant's cardiorespiratory status by watching their work and rate of breathing. The overall quality of this assessment is often superior in the home environment: the infant is frequently calm, interacting with parent in a deeply personal manner which is strikingly different from the exam that the team is able to obtain in clinic after the family has been through prolonged travel, the stress of a physical exam and an echocardiogram. There are now over 17,000 videos in our database.

Recent work at CMKC seeks to analyze the video data to determine if it could be analyzed in a manner akin to the Cardiac-Early Warning Score (CHEWS), which is used to predict clinical decompensation and

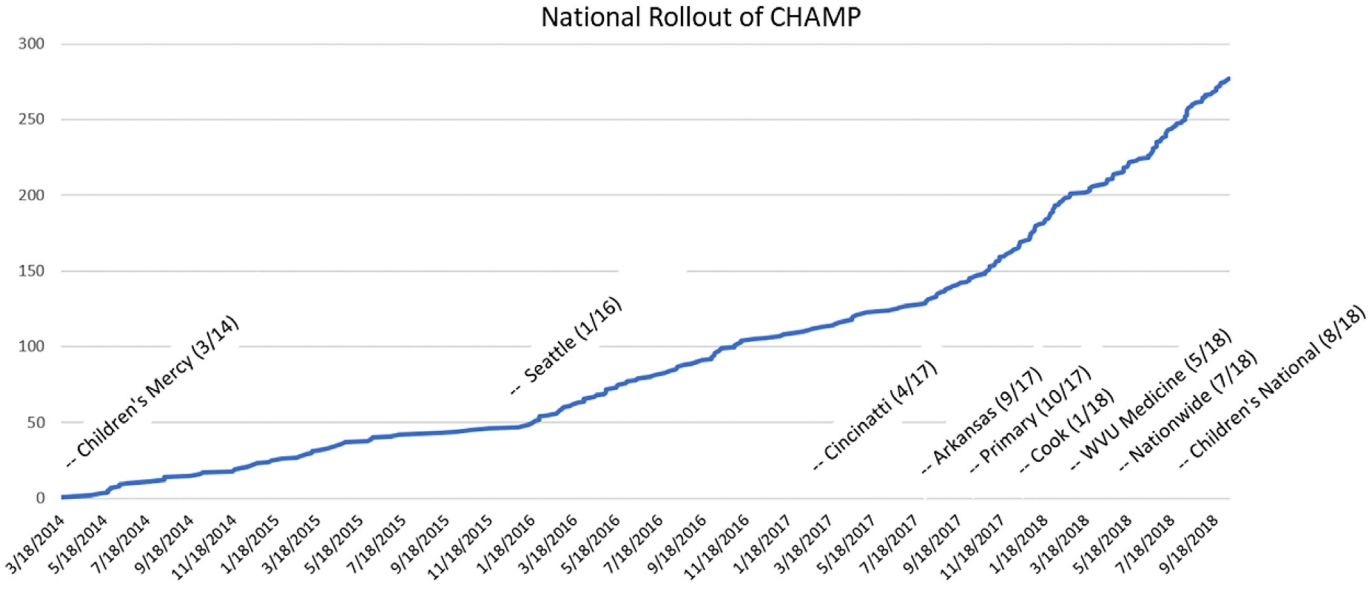

Fig. 2. Dates of each participating site with total number of patients registered (blue line). The past year has seen our total participating sites rise to 8.

unplanned hospital admission [19–22]. We have scored videos based on observed respiratory rate, respiratory effort, color, behavior and general appearance, with higher scores suggesting greater clinical risk. At CMKC, there were 64 unplanned admissions since CHAMP was initiated. Baseline videos (> 14 days before re-admission) were scored compared to case videos (within 48 h of unplanned admission). Scoring was feasible for 92% of variables and was highly reproducible between observers. The case videos had much higher scores than baseline (6.9 versus 1.7, $p < 0.001$) suggesting that this mode of quantifying information from CHAMP videos may provide objective warning of acute decompression.

9. Future directions

While we are pleased with our progress thus far, there are ongoing opportunities for this technology to assist in the lives of this patient population. We plan to continue to expand to additional partner sites. We are in the process of rolling out a mobile platform (iOS and Android apps) as our parents and families evolve to an increasingly mobile environment. We are continuing our work towards predictive analytics using combinations of CHAMP tablet data and patient-specific EMR-based data. We hope that our video analysis could be replicated and predictive across our partner sites.

It became clear to our team several years ago that every patient who is enrolled in CHAMP is contributing to a live registry. The large amount of home-based data, combined with outcome-based data from the electronic health record in the CHAMP database presents unique opportunities for clinical research. We are optimistic that the CHAMP registry could eventually serve as a platform to base a randomized-registry type trial, at a fraction of the cost of a traditional randomized trial [23].

10. Summary

CHAMP serves as a model to envision a different paradigm of care for our fragile interstage patients. While some of the gaps with the prior model of care were evident at the outset (data loss, data burden for the parent), some of the advantages of the new model have only become apparent with experience (minimizing delays in care, benefit of videos, equity among patients). There were also some differences between intent and reality; for example, we now have increased data entry burden for the care teams. Overall, the largest transformation is that we are following these patients at home in a proactive, real-time manner which allows quicker response and intervention before a potential clinical decompensation. Thus, CHAMP represents a new paradigm, providing for a continuum of monitoring and care from inpatient to outpatient care. We are optimistic that we will continue to improve the system through an iterative process, and to continue to reduce morbidity and mortality for this fragile group.

Declaration of Competing Interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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