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Abstract

Objective: Patients after Fontan operation for complex congenital heart disease (CHD) have decreased exercise capacity and report reduced health-related quality of life (HRQOL). Studies suggest hospital-based cardiac physical activity programs can improve HRQOL and exercise capacity in patients with CHD; however, these programs have variable adherence rates. The impact of a home-based cardiac physical activity program in Fontan survivors is unclear. This pilot study evaluated the safety, feasibility, and benefits of an innovative home-based physical activity program on HRQOL in Fontan patients.

Methods: A total of 14 children, 8–12 years, with Fontan circulation enrolled in a 12-week moderate/high intensity home-based cardiac physical activity program, which included a home exercise routine and 3 formalized in-person exercise sessions at 0, 6, and 12 weeks. Subjects and parents completed validated questionnaires to assess HRQOL. The Shuttle Test Run was used to measure exercise capacity. A Fitbit Flex Activity Monitor was used to assess adherence to the home activity program.

Results: Of the 14 patients, 57% were male and 36% had a dominant left ventricle. Overall, 93% completed the program. There were no adverse events. Parents reported significant improvement in their child's overall HRQOL ($P < .01$), physical function ($P < .01$), school function ($P = .01$), and psychosocial function ($P < .01$). Patients reported no improvement in HRQOL. Exercise capacity, measured by total shuttles and exercise time in the Shuttle Test Run and calculated $VO_2\max$, improved progressively from baseline to the 6 and 12 week follow up sessions. Monthly Fitbit data suggested adherence to the program.

Conclusion: This 12-week home-based cardiac physical activity program is safe and feasible in preteen Fontan patients. Parent proxy-reported HRQOL and objective measures of exercise capacity significantly improved. A 6-month follow up session is scheduled to assess sustainability. A larger study is needed to determine the applicability and reproducibility of these findings in other age groups and forms of complex CHD.

Introduction

The Fontan operation, typically performed between 2 and 4 years of age, is a palliative procedure for patients born with complex congenital heart disease (CHD) characterized by a functional single ventricle. The operation diverts the systemic venous return directly to the pulmonary arteries, resulting in a passive, nonpulsatile pulmonary circulation. Despite improved survival, patients after the Fontan operation report reduced health-related quality of life (HRQOL), particularly in the domain of physical functioning,¹⁻⁴ and have decreased exercise tolerance,⁵⁻⁷ compared to peers.

The etiology of reduced physical function HRQOL in Fontan patients is likely multifactorial. Previously identified risk factors include reduced exercise capacity, residual hemodynamic defects, chronotropic impairment, deconditioning secondary to physical inactivity, and psychosocial factors such as parental overprotection and/or social restraints.⁸ Cardiac rehabilitation programs, which incorporate structured exercise training programs, are successful in improving patient-reported HRQOL and objective measurements of exercise capacity in adult patients with acquired heart disease.⁹ Similarly, a small number of studies have suggested that hospital-based cardiac physical activity programs may result in similar improvements in children and young adults with complex CHD.⁹⁻¹⁵ These results suggest that physical function HRQOL is modifiable and may improve with programs that encourage an increase in physical activity and exercise. However, participation in hospital-based programs have variable

adherence rates due to inaccessibility and inconvenience for pediatric patients, who rely on family members limited by work commitments, finances, location, and/or transportation.

More recent studies have indicated a home-based physical activity program, specifically tailored to pediatric patients, may be more accessible to patients and can improve HRQOL in overweight and obese children.¹⁶ However, the impact of a home-based physical activity program on the physical function HRQOL in patients with Fontan circulation is unclear.

The purpose of this study was to determine the safety, feasibility, and benefits of an innovative 12-week home-based cardiac physical activity program on the physical function HRQOL, as well as measurements of exercise capacity in children, 8–12-years-old, with Fontan circulation.

Methods

The study was approved by the Children's Hospital of Wisconsin Internal Human Research Review Board and was conducted in accordance with all human research regulatory requirements.

Patients

We recruited 8- to 12-year-old patients with Fontan physiology, followed in the Children's Hospital of Wisconsin Pediatric Cardiology Clinic, for enrollment in a 12-week home-based physical activity program. The age range of 8–12 years was chosen based on the validation studies for the HRQOL questionnaires used for this study. Exclusion criteria included indications for restriction from exercise activity of greater than moderate intensity based on American Heart Association recommendations.¹⁷ These include: (1) moderate or severe cardiac systolic dysfunction based on echocardiography or cardiac magnetic resonance imaging study within 12 months of study enrollment, (2) aortic dilation with ascending aorta dimension >4 cm, (3) history of exertional syncope, and (4) history of atrial and/or ventricular arrhythmias. In addition, patients with a noncardiac medical or psychiatric disorder that would prevent successful

completion of planned study testing or would invalidate the results of the study testing were excluded.¹⁷ None of the patients had a pacemaker or implantable cardioverter-defibrillator. Patients were included in the study once informed assent of the study participant and consent of the parent or guardian were obtained, according to institutional guidelines.

Physical Activity Program

The 12-week home-based moderate-to-high intensity physical activity program consisted of 2 primary components: (1) a 45-minute home exercise routine of dynamic and static exercises, depicted on DVD or paper handout, for subjects to complete 3–4 times per week and (2) 3 formalized in-person exercise sessions.

Assessments of physical function HRQOL were made with the Pediatric Quality of Life Inventory (PedsQL, Mapi Research Trust, Lyon, France). The child and parent-proxy report for the PedsQL questionnaire has been validated in children 8–12-years-old and for parent proxy-reporting.¹⁸⁻²⁰ The questionnaires were administered prior to initiation of the physical activity program (baseline) and at the end of the 12-week physical activity program.

Exercise capacity was assessed with exercise duration and estimates of maximal oxygen uptake ($VO_2\text{max}$) based on performance on the 20-meter Shuttle Test Run (Personal Fitness Tests, Poole, United Kingdom).²¹ Participants were asked to run a 20-meter fixed distance at increasing workloads (speeds) using a standard prerecorded audio CD. Testing was terminated once a patient could no longer maintain the prescribed pace. The total number of shuttles (TL) the participant was able to achieve was recorded and the estimated $VO_2\text{max}$ could then be calculated using a validated equation:
 $VO_2\text{max} = 61.1 - 2.20(\text{gender: M} = 0; \text{F} = 1) - 0.462(\text{age}) - 0.862(\text{BMI}) + 0.192(\text{TL}).$ ²²⁻²⁵

Additional activity during the 12-week study period was not limited and participants were encouraged to be as active and he/she desired. To assess and document adherence to the home exercise protocol, subjects recorded their daily activity in a journal and reported their activity log during a scheduled telephone call. In

addition, each participant was given their own FitBit Flex (Fitbit; San Francisco, CA, USA) Activity Monitor, to objectively measure physical activity and adherence during the 12-week physical activity program. The Fitbit Flex Activity Monitor is an accelerometer that is worn like a small wristband and records data on movement (steps, distance, and calories), total activity minutes, and total minutes of inactivity and sleep. The data is stored on the device and then can be downloaded wirelessly to a computer. Each participant was invited to join a secure online group through the FitBit website to evaluate his/her performance compared to the other study participants. In addition, study participants and their parents were able to network their questions and concerns with each other through this secure group. A monthly group progress report could be accessed and printed to evaluate and compare each participant's activity level throughout the study period.

Data Outcomes and Risk Factors

Each participant's baseline height, weight, and BMI were recorded for the first and third in-person session. In addition, each participant's resting and immediately post Shuttle Test Run oxygen saturation, blood pressure, and heart rate were recorded. Retrospective chart review was performed to identify relevant clinical and demographic information for the study subjects, including results of their most recent transthoracic echocardiogram and cardiopulmonary exercise test. Systemic ventricular systolic function was assessed semiquantitatively by a single pediatric cardiologist, and graded as normal, mildly diminished, moderately diminished, or severely diminished. Adverse events were also recorded during the study period, including the development of arrhythmias, bodily injury, need for hospitalization, cardiac arrest, and death.

Statistical Analysis

Descriptive statistics, mean with standard deviation/s or median with ranges, were used to summarize the demographic characteristics of study participants. The primary outcome measure in our study was the physical function PedsQL scores, calculated at the beginning of the intervention and immediately after the 12-week intervention. The

secondary outcome measure was change in exercise capacity as measured by shuttle number, time, and distance, and the calculated VO₂max assessed with the Shuttle Test Run during the same time periods. Other physical parameters of height, weight, BMI, and resting and immediately postexercise heart rate, blood pressure, and oxygen saturation were also analyzed.

Physiologic parameters are presented as mean with standard deviation. Paired sample two-tailed Student *t*-tests were performed to compare differences between pre- and postintervention outcomes. Additionally, a repeated measure ANOVA was performed to compare mean differences between each of the 3 time points within the study (baseline, 6 weeks, and 12 weeks). Post hoc analysis using the least significant differences (LSD) method was used to determine differences. Linear regression was performed to assess the contributions of independent variables toward outcome variables. Pearson correlations were calculated to determine if relationships existed between physiologic outcomes. Questionnaire data were scored and totaled per instrument standards. Total scores from the questionnaires were compared pre- to postrehabilitation using Student *t*-tests. If violations of parametric data occurred, nonparametric assessment of differences in pre-post values was established via Wilcoxin signed-rank test. All significance was established at $P < .05$.

Results

The demographics and baseline characteristics for the study cohort are summarized in Table 1. Of the 14 patients in our study, the median age was 10 years (range, 8–12 years) and 8 (57%) were male. Most of the patients had a dominant right ventricle (8 [57%]), followed by 5 patients (36%) with a dominant left ventricle, and 1 patient (7%) had biventricular anatomy, with mitral valve atresia, mild left ventricular hypoplasia, and a straddling right atrioventricular valve. Twelve patients (86%) had an extracardiac Fontan and 2 patients (14%) had a lateral tunnel Fontan. Seven patients (50%) had a patent fenestration. One patient had a previous diagnosis of plastic bronchitis. In our study, there were 8 patients (57%) who had neurocognitive, neurologic, or psychiatric comorbidities, including anxiety disorder in 3 patients, attention-deficit disorder in 3 patients,

developmental disability in 2 patients, and a seizure disorder in 1 patient.

Table 1. Baseline Patient Demographics

| ID | Age | BMI (kg/m ²) | CHD Diagnosis | Dominant Ventricle | Fenestration | Time since Fontan | ECHO Function |
|----|-----|-----------------------------|--|-----------------------|--------------|-------------------------|-------------------|
| 1 | 9 | 14.2 | DORV w/mitral hypoplasia | RV | No | 6.8 yrs | Normal |
| 2 | 11 | 18.6 | TA | LV | Yes | 8.3 yrs | Normal |
| 3 | 10 | 17.7 | TA | LV | No | 5.4 yrs | Normal |
| 4 | 10 | 14.5 | HLHS | RV | No | 4.5 yrs | Normal |
| 5 | 12 | 15.8 | HRHS | LV | Yes | 8.5 yrs | Normal |
| 6 | 12 | 20.1 | HLHS | RV | Yes | 9.2 yrs | Normal |
| 7 | 9 | 14 | TA | LV | No | 6.9 yrs | Normal |
| 8 | 9 | 22 | Unbalanced AVSD | RV | Yes | 1.7 yrs | Normal |
| 9 | 9 | 16.4 | Coarctation with mitral & LV hypoplasia | RV | No | 6.8 yrs | Mildly diminished |
| 10 | 9 | 16.3 | HLHS | RV | Yes | 7.3 yrs | Normal |
| 11 | 12 | 23.3 | TA | LV | No | 11.1 yrs | Normal |
| 12 | 12 | 15.3 | Unbalanced AVSD | RV | No | 9.5 yrs | Normal |
| 13 | 10 | 16.4 | D-TGA w/mitral atresia & straddling right AV valve | LV | Yes | 7.1 yrs | Normal |
| 14 | 12 | 23.6 | HLHS | RV | Yes | 9.9 yrs | Normal |

Based on the most recent echocardiogram, all of the patients were considered to have normal or mildly diminished systemic ventricular systolic function. In addition, all of the patients had completed a treadmill cardiopulmonary exercise test within 25 months of the first in-person session (range, 1–25 months). However, only 29% (4/13) could cooperate for metabolic testing and measurement of VO₂max by breath-by-breath analysis during the cardiopulmonary exercise testing. For these 4 patients, the timing of the cardiopulmonary testing was variable relative to the initiation of the study. Therefore, an accurate comparison of each participant's VO₂max

achieved on the cardiopulmonary stress test and that calculated from the Shuttle Test Run could not be performed.

Eleven of the participants lived within approximately 60 miles of the hospital (range, 4–46 miles); however, there were 3 families who had to travel a greater distance (range, 93–121 miles) to attend the 3 in-person sessions. Ten (71%) of the patients reported themselves as physically active prior to the study.

Ninety-three percent (13/14) of the subjects completed the 12-week program. There were no adverse events during the study period. One patient was unable to continue participation after the first in-person session, due to personal family issues unrelated to the study. Adherence was excellent. All of the 13 participants who completed the study turned in their daily activity log at each of the 3 in-person sessions. The activity logs were similar to the Fitbit Flex Activity Monitor data (steps, total distance, active minutes) available through a secure online community in 10/13 patients. The Fitbit data over the 12-week study period is shown in Figure 1.

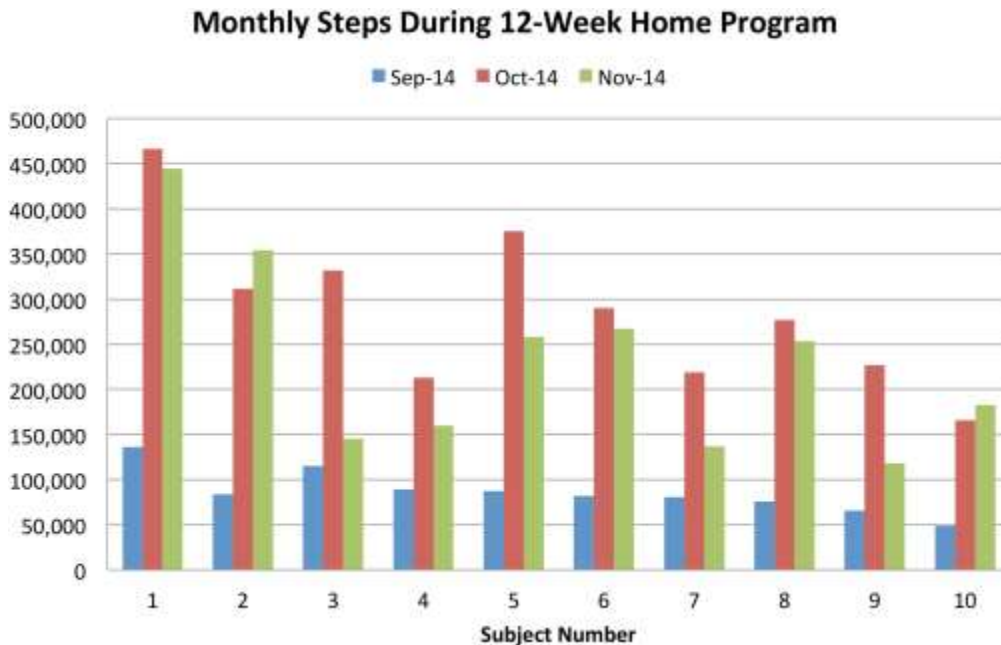


Figure 1. Fitbit Activity Monitor data over a 12-week physical activity program. Only 10 of the 14 patients chose to participate in the online Fitbit activity monitoring.

There were 2 patients who never did sign up to be a part of the online private community and consistently forgot to print off their

information to turn in with their activity log. In addition, 1 patient lost their Fitbit and had to replace it halfway through the study.

There was no measurable improvement in the patient-reported HRQOL after completion of the 12-week program. However, parents reported significant improvement in their child's overall HRQOL ($P = .004$), physical function ($P = .006$), school function ($P = .01$), social function ($P = .005$), and psychosocial function ($P = .005$) on the PedsQL (Table 2).

Table 2. PedsQL Mean 4.0 Generic Scores: Comparison of Child Self-report and Parent Proxy-report at Baseline and Following Completion of 12-Week Physical Activity Study

| | n | Pre Mean ± SD | n | Post Mean ± SD | Pre vs. Post P value |
|---|----|------------------|----|-------------------|-------------------------|
| 1. The bold numbers represent the statistically significant P value between pre- and post-PedsQL score. | | | | | |
| Children | | | | | |
| PedsQL | | | | | |
| Overall | 13 | 73.8 ± 10.9 | 13 | 74.2 ± 7.10 | 0.890 |
| Physical | 13 | 74.8 ± 11.4 | 13 | 75.0 ± 9.63 | 0.947 |
| Emotional | 13 | 75.0 ± 19.6 | 13 | 74.2 ± 13.9 | 0.814 |
| Social | 13 | 78.5 ± 13.6 | 13 | 81.5 ± 6.25 | 0.464 |
| School | 13 | 66.5 ± 11.8 | 13 | 64.4 ± 15.1 | 0.781 |
| Psychosocial | 13 | 73.3 ± 12.8 | 13 | 73.7 ± 9.6 | 0.892 |
| Parent | | | | | |
| PedsQL | | | | | |
| Overall | 13 | 63.8 ± 13.7 | 13 | 73.5 ± 12.3 | 0.004 |
| Physical | 13 | 70.7 ± 13.4 | 13 | 78.8 ± 14.6 | 0.006 |
| Emotional | 13 | 63.5 ± 19.1 | 13 | 70.8 ± 21.5 | 0.145 |
| Social | 13 | 62.7 ± 24.1 | 13 | 71.5 ± 20.1 | 0.005 |
| School | 13 | 54.2 ± 16.2 | 13 | 68.8 ± 14.7 | 0.01 |
| Psychosocial | 13 | 60.1 ± 16.1 | 13 | 70.4 ± 12.5 | 0.005 |

There was no statistically significant change in the participants' resting and post exercise vital signs, including heart rate, oxygen saturation, and blood pressure, from the initial session to the end of the 12-week study.

Exercise capacity, measured by the number of shuttles, distance, and time completed in the 20-meter Shuttle Test Run improved significantly from baseline to the 12-week session. The overall mean shuttle time for the group improved from baseline to the

6-week session ($P = .003$). Per patient, there was a mean percentage improvement in total exercise time of 22% at the 6-week session. In addition, the overall mean shuttle time for the group improved from baseline to the 12-week session ($P = .01$), with a per-patient mean percentage increase of 24% (Figure 2). There was also a statistically significant change in calculated VO_2 max from baseline to the 12-week session, improving from a mean of 41.8 to 42.3 mL/kg/min ($P = .001$) (Figure 3).

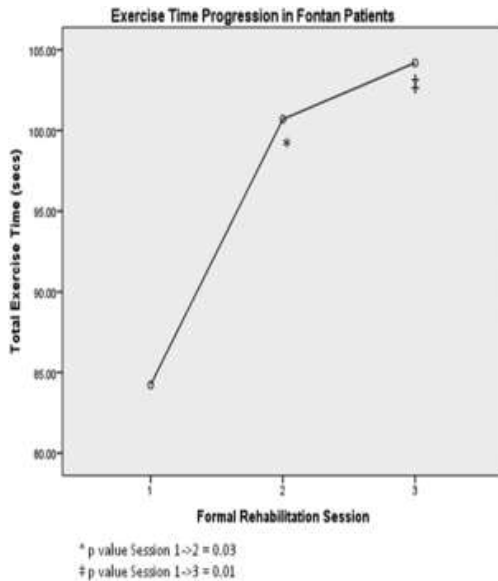


Figure 2. Change in exercise time on the 20-meter Shuttle Test Run from baseline to completion of 12-week physical activity.

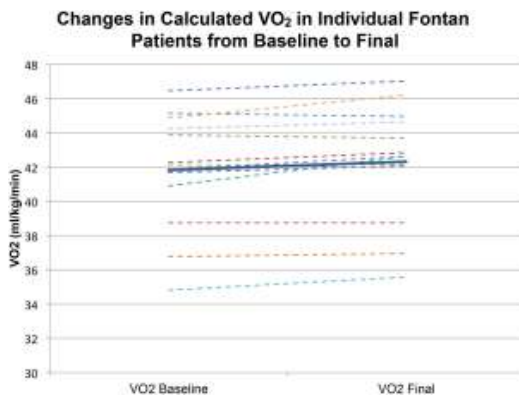


Figure 3. Changes in calculated VO_2 max throughout a 12-week physical activity program. VO_2 , peak oxygen consumption. Each dotted line represents a single participant and the solid line the overall group, with each line representing the mean change in peak oxygen consumption from baseline to completion of the 12-week study.

On linear regression analysis, there was no association between any of the patients' clinical variables and outcome measures of HRQOL and exercise capacity.

Discussion

In this study, we demonstrated a 12-week moderate-to-high intensity home-based cardiac physical activity program is safe and feasible for children with Fontan circulation. In addition, there were significant improvements in parents' reports of their child's physical function HRQOL as well as objective measurements of the participants' exercise capacity. However, the program did not significantly improve patients' report of their own physical function HRQOL.

The improvement in parental perceptions of their child's HRQOL is potentially a clinically relevant outcome. Previous studies have shown that parental anxiety about the safety of physical activity, as well as inadequate physician-parent communication and/or agreement on physical activity restrictions, likely contribute to the reduced physical function HRQOL reported by parents of children after the Fontan procedure.^{8,26} An intervention, such as our physical activity program, which can improve parental perceptions of their child's capabilities, may promote increased physical activity in this vulnerable population.

The reasons for the lack of improvement in patients' self-reported physical function HRQOL after participation in the 12-week program, despite improvements in objective measures of exercise capacity, are unclear. One hypothesis is that the children in our volunteer study already had a high perception of their physical function HRQOL, making it difficult to detect a significant increase with a pilot study of only 14 patients. Another hypothesis is that exercise capacity has less influence on physical function HRQOL compared to other important clinical factors. In fact, recent studies demonstrate that objective measures of cardiac function and exercise capacity do not correlate, or only weakly correlate, with self-reported HRQOL in patients with Fontan circulation.^{14,27} In the Pediatric Heart Network multicenter Fontan study, noncardiac problems had the greatest impact on physical function HRQOL scores.¹⁴

Previous single-center studies have also demonstrated benefits from a cardiac physical activity program. Rhodes et al. reported improvements in objective measurements of exercise capacity and scores for emotional, behavioral, and physical function HRQOL, assessed with a patient questionnaire, after a 12-week hospital-based cardiac rehabilitation program in 15 patients with complex CHD (including 10 patients with Fontan circulation). Importantly, the improvement in exercise capacity as well as HRQOL was sustained at 1 year follow up; whereas the HRQOL scores for patients with CHD in the control group who did not undergo rehabilitation actually declined.^{10,11} Similarly, Dua et al. also demonstrated that an exercise-training program significantly improved the physical self-perception, life satisfaction, physical activity levels, and general health in a cohort of CHD patients, including those with Fontan circulation.¹²

Our study differs from the studies by Rhodes et al. and Dua et al. in that the majority of participation in our study was home-based. In a recent study, Longmuir et al. reported their results from a home-based physical activity program in children with Fontan circulation, 6–12 years old, over a 2-year time period. Patients were randomized into an activity education versus activity prescription cohort. Similar to our experience, there were no adverse events and the majority of children successfully completed the program. At the end of study, they demonstrated both activity education and activity prescription are equally effective for maintaining moderate-to-vigorous physical activity and improving gross motor skill, exercise capacity, and fitness. Interestingly, there was no change in activity attitudes, as assessed through verbal administration of Children's Self-Perception of Adequacy & Predilection for Physical Activity Scale.²⁸

Collectively, the results from our study and the previously discussed studies suggest that physical function HRQOL and exercise capacity is modifiable in patients with Fontan circulation and may improve with programs that encourage an increase in physical activity and exercise. Our study is unique in that the majority of participation was home-based, and thus the program could be completed by patients that reside long distances from our hospital center. In addition, it demonstrated improvement in parent-proxy HRQOL, particularly in the domain of physical function, as well as exercise capacity.

Limitations

Our study does have several limitations. This was a pilot study limited by small size, at a specific age range, and the subjects and families who volunteered for the study were likely highly motivated, thus limiting the generalizability to other patients with Fontan circulation. The measurements of exercise capacity obtained with the Shuttle Test Run have been validated to calculate VO_2 max in normal children, but it has not been validated for children with CHD. Our intention was to compare the VO_2 max from each participant's treadmill cardiopulmonary exercise test to his/her calculated VO_2 max from the Shuttle Test Run. However, the majority of children did not have a cardiopulmonary exercise test with VO_2 max measured by breath-by-breath analysis within an acceptable time to the start of our study. Although a statistically significant improvement in VO_2 max was detected, it is difficult to quantify the clinical relevance of this change. In addition, while useful, the Fitbit Flex Activity Monitor is new technology, and as a result, there were technical difficulties as our families learned how to use them.

Conclusion

A home-based cardiac physical activity program is safe and feasible in children who have undergone a Fontan procedure. In addition, parent proxy-reported HRQOL and objective measures of exercise capacity improved significantly following the 12-week program. A 6-month follow up session is scheduled to assess the sustainability of the improvements in HRQOL and a physically active lifestyle. A larger, randomized study is needed to determine the applicability and reproducibility of these findings in other age groups of Fontan survivors and other forms of complex CHD.

Author Contributions

Roni M. Jacobsen, MD: Concept/design, data collection, data analysis/interpretation, drafting article, approval of article; Salil Ginde, MD: Concept/design, data collection, data analysis/interpretation, drafting article, approval of article; Kathleen Mussatto, PhD, RN: Concept/design, data collection, data analysis/interpretation, drafting article, approval of article; Jennifer Neubauer, RN, MSN, CPNP: Data collection, data analysis/interpretation, drafting article, approval of article; Michael Earing,

MD: Concept/design, data analysis/interpretation, drafting article, approval of article; Michael Danduran, MS, RCEP-ACSM Concept/design, data collection, statistics, data analysis/interpretation, drafting article, approval of article

References

- ¹ Muller J, Christov F, Schreiber C, Hess J, Hager A. Exercise capacity, quality of life, and daily activity in the long-term follow-up of patients with univentricular heart and total cavopulmonary connection. *Eur Heart J*. 2009;30(23):2915–2920.
- ² Manlihot C, Knezevich S, Radojewski E, Cullen-Dean G, Williams WG, McCrindle BW. Functional health status of adolescents after the fontan procedure—comparison with their siblings. *Can J Cardiol*. 2009;25(9):e294–e300.
- ³ Uzark K, Jones K, Slusher J, Limbers CA, Burwinkle TM, Varni JW. Quality of life in children with heart disease as perceived by children and parents. *Pediatrics*. 2008;121(5):e1060–e1067.
- ⁴ McCrindle BW, Williams RV, Mitchell PD, et al. Relationship of patient and medical characteristics to health status in children and adolescents after the fontan procedure. *Circulation*. 2006;113(8):1123–1129.
- ⁵ Giannico S, Hammad F, Amodeo A, et al. Clinical outcome of 193 extracardiac fontan patients: the first 15 years. *J Am Coll Cardiol*. 2006;47(10):2065–2073.
- ⁶ Driscoll DJ. Cardiorespiratory responses to exercise after the fontan operation. *Circulation*. 1990;81(6):2016–2017.
- ⁷ Paridon SM, Mitchell PD, Colan SD, et al. A cross-sectional study of exercise performance during the first 2 decades of life after the fontan operation. *J Am Coll Cardiol*. 2008;52(2):99–107.
- ⁸ Longmuir PE, McCrindle BW. Physical activity restrictions for children after the fontan operation: disagreement between parent, cardiologist, and medical record reports. *Am Heart J*. 2009;157(5):853–859.
- ⁹ Taylor RS, Brown A, Ebrahim S, et al. Exercise-based rehabilitation for patients with coronary heart disease: systematic review and meta-analysis of randomized controlled trials. *Am J Med*. 2004;116(10):682–692.
- ¹⁰ Rhodes J, Curran TJ, Camil L, et al. Impact of cardiac rehabilitation on the exercise function of children with serious congenital heart disease. *Pediatrics*. 2005;116(6):1339–1345.
- ¹¹ Rhodes J, Curran TJ, Camil L, et al. Sustained effects of cardiac rehabilitation in children with serious congenital heart disease. *Pediatrics*. 2006;118(3):e586–e593.
- ¹² Dua JS, Cooper AR, Fox KR, Graham Stuart A. Exercise training in adults with congenital heart disease: feasibility and benefits. *Int J Cardiol*. 2010;138(2):196–205.

- 13 Longmuir PE, Russell JL, Corey M, Faulkner G, McCrindle BW. Factors associated with the physical activity level of children who have the fontan procedure. *Am Heart J.* 2011;161(2):411–417.
- 14 McCrindle BW, Williams RV, Mital S, et al. Physical activity levels in children and adolescents are reduced after the fontan procedure, independent of exercise capacity, and are associated with lower perceived general health. *Arch Dis Child.* 2007;92(6):509–514.
- 15 Ewalt LA, Danduran MJ, Strath SJ, Moerchen V, Swartz AM. Objectively assessed physical activity and sedentary behavior does not differ between children and adolescents with and without a congenital heart defect: a pilot examination. *Cardiol Young.* 2012;22(1):34–41.
- 16 Lison JF, Real-Montes JM, Torro I, et al. Exercise intervention in childhood obesity: a randomized controlled trial comparing hospital-versus home-based groups. *Acad Pediatr.* 2012;12(4):319–325.
- 17 Longmuir PE, Brothers JA, de Ferranti SD, et al. Promotion of physical activity for children and adults with congenital heart disease: a scientific statement from the American Heart Association. *Circulation.* 2013;127(21):2147–2159.
- 18 Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the pediatric quality of life inventory version 4.0 generic core scales in healthy and patient populations. *Med Care.* 2001;39(8):800–812.
- 19 Varni JW, Seid M, Rode CA. The PedsQL: measurement model for the pediatric quality of life inventory. *Med Care.* 1999;37(2):126–139.
- 20 Varni JW, Burwinkle TM, Seid M, Skarr D. The PedsQL 4.0 as a pediatric population health measure: feasibility, reliability, and validity. *Ambul Pediatr.* 2003;3(6):329–341.
- 21 Personal Fitness Tests. (2011). *The Bleep Test Training Program.* Poole, UK.
- 22 Matsuzaka A, Takahashi Y, Yamazoe M, et al. Validity of the multistage 20-m shuttle-run test for Japanese children, adolescents, and adults. *Pediatr Exerc Sci.* 2004;16:113–25.
- 23 Mahar, MT, Guerieri, AM, Hanna, MS, Kembles, CD. Estimation of aerobic fitness from 20-m multistage shuttle run test performance. *Am J Prev Med.* 2011;41(4s2):S111–S123.
- 24 Leger LA, Mercier D, Gadoury C, Lambert J. The multistage 20 metre shuttle run test for aerobic fitness. *J Sports Sci.* 1988;6(2):93–101.
- 25 Barnett A, Chan LYS, Bruce IC. A preliminary study of the 20-m multistage shuttle run as a predictor of a peak VO₂ in Hong Kong Chinese students. *Pediatr Exerc Sci.* 1993;5:42–50.
- 26 Moola F, Fusco C, Kirsh JA. The perceptions of caregivers toward physical activity and health in youth with congenital heart disease. *Qual Health Res.* 2011;21(2):278–291.
- 27 Burns R, Olson I, Kazmucha J, Balise R, Chin R, Chin C. Correlation of subjective questionnaires with cardiac function as determined by

exercise testing in a pediatric population. *Pediatr Cardiol.* 2010;31(7):1043–1048.

- ²⁸ Longmuir PE, Tyrrell PN, Corey M, et al. Home-based rehabilitation enhances daily physical activity and motor skill in children who have undergone the fontan procedure. *Pediatr Cardiol.* 2013;34(5):1130–1151.

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