

# UC Irvine

## UC Irvine Previously Published Works

### Title

Optimism despite profound uncertainty: school and social relationships in adolescents with single ventricle heart disease.

### Permalink

<https://escholarship.org/uc/item/9q86d56g>

### Journal

Cardiology in the young, 31(2)

### ISSN

1047-9511

### Authors

Peterson, Jennifer K  
Olshansky, Ellen F  
Guo, Yuqing  
[et al.](#)

### Publication Date

2021-02-01

### DOI

10.1017/s1047951120003790

Peer reviewed



Published in final edited form as:

*Cardiol Young*. 2021 February ; 31(2): 252–259. doi:10.1017/S1047951120003790.

## Optimism Despite Profound Uncertainty: School and Social Relationships in Adolescents with Single Ventricle Heart Disease

**Jennifer K. Peterson,**

Assistant Professor, Johns Hopkins University School of Nursing, Baltimore, MD

**Ellen F. Olshansky,**

Professor Emerita, Sue and Bill Gross School of Nursing, University of California, Irvine, Irvine, CA

**Yuqing Guo,**

Associate Professor Sue and Bill Gross School of Nursing, University of California, Irvine, Irvine, CA

**Lorraine S. Evangelista,**

Associate Dean for Research & Scholarship and Professor University of Texas Medical Branch School of Nursing

**Nancy A. Pike**

Associate Professor, School of Nursing, University of California, Los Angeles;

Nurse Practitioner, Cardiothoracic Surgery, Children's Hospital, Los Angeles

### Abstract

**Background:** Survivors of single ventricle heart disease must cope with the physical, neurodevelopmental, and psychosocial sequelae of their cardiac disease, which may also affect academic achievement and social relationships. The purpose of this study was to qualitatively examine the experiences of school and social relationships in adolescents with single ventricle heart disease.

**Methods:** A descriptive phenomenological methodology was employed, utilizing semi-structured interviews. Demographic and clinical characteristics were obtained via chart review.

**Results:** Fourteen adolescents (ages 14 to 19 years) with single ventricle heart disease participated. Interviews ranged from 25 to 80 minutes in duration. Four themes emerged from the interviews, including 1) “Don’t assume”: Pervasive ableism; 2) “The elephant in the room”: Uncertain future; 3) “Everyone finds something to pick on”: Bullying at school; 4) “They know what I have been through”: Social support. The overall essence generated from the data was “optimism despite profound uncertainty.”

---

**Corresponding Author:** Jennifer K. Peterson, Ph.D., APRN-CNS, CCNS, Johns Hopkins University School of Nursing, 525 North Wolfe St, Room 460, Baltimore, MD 21205, [jpete107@jhu.edu](mailto:jpete107@jhu.edu), Telephone 714-280-3219.

**Conflicts of Interest:** All authors have no financial or other conflicts of interest

**Ethical Standards:** The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant guidelines on human experimentation (United States of America) and has been approved by the institutional committee at the University of California, Los Angeles.

**Conclusions:** Adolescents with single ventricle heart disease identified physical limitations and school challenges in the face of an uncertain health-related future. Despite physical and psychosocial limitations, most remained optimistic for the future and found activities that were congruent with their abilities. These experiences reflect “optimism despite profound uncertainty.”

### Keywords

Fontan Procedure; Qualitative Research; Academic Achievement; Peer Relationships

---

### Introduction

Improved survival in patients with congenital heart disease (CHD) has led to a growing population of adolescents and young adults with single ventricle heart disease who have undergone Fontan palliation.<sup>1,2</sup> However, the inherently abnormal Fontan physiology is often associated with long-term medical and end-organ complications including heart failure, arrhythmias, chronic hepatic injury, protein-losing enteropathy, and other lymphatic-related diseases such as plastic bronchitis.<sup>3–5</sup> Varying degrees of neurodevelopmental and cognitive delays are common in Fontan survivors and are assumed to be multifactorial in origin.<sup>5,6</sup> These complications may adversely impact mortality and morbidity as well as physical functioning, quality of life, academic achievement, social relationships, and ability for self-care.

This “neurodevelopmental phenotype” of complex CHD includes mild cognitive delays, motor delays, impaired social cognition and communication skills, impaired executive functioning, and behavioral problems, including attention deficit hyperactivity disorder and autism spectrum disorder.<sup>7</sup> Anxiety and depression are common mental health concerns identified in adolescents with single ventricle heart disease.<sup>8,9</sup> These neurodevelopmental and psychological co-morbidities can evolve, and new problems may emerge as developmental and functional demands increase.<sup>10</sup> Academic challenges in adolescents with single ventricle heart disease include lower scores on tests of reading and mathematics than controls<sup>11</sup> as well as higher utilization of special education services.<sup>12</sup> Adolescents with chronic health problems are also at increased risk for bullying, often related to being perceived as different from their peers.<sup>13</sup> The effect of these neurodevelopmental and psychosocial sequelae on the ability to form stable and healthy social relationships is not well studied, but impairments in social cognition may contribute to impaired psychosocial functioning.<sup>14,15</sup>

These many challenges form the context in which adolescents with single ventricle heart disease experience school and social relationships, which also form the basis for achieving developmental milestones that result in career success and independent living.<sup>15,16</sup> Despite this, little is known qualitatively about school experiences and social relationships in this at-risk population. The purpose of this study was to qualitatively explore the daily lives of Fontan survivors to gain a better understanding of how single ventricle heart disease affected their experiences of school and social relationships, to identify potential support strategies.

## Materials and Methods

### Design

This qualitative study employed a descriptive phenomenological methodology, described by Edmund Husserl.<sup>17</sup> A phenomenological approach is well-suited to identify the meaning, structure, and essence of the experiences of a particular phenomenon for a person or group of people.<sup>18,19</sup> The approach of descriptive phenomenology is to identify the experience of interest, bracket out or set aside the influence of one's own experiences, and collect data from those who have lived the experience.<sup>19,20</sup> Data analysis follows steps outlined in the data analysis section. This study was approved by the University of California, Los Angeles Institutional Review Board (IRB).

### Sample and Recruitment

Adolescents aged 14 to 19 years with single ventricle heart disease who had undergone Fontan completion were purposively sampled. Potential participants were recruited from a larger single ventricle heart disease cohort study evaluating cognitive outcomes and brain structural integrity<sup>8,21</sup>, who had indicated on the informed consent a willingness to be contacted for future studies. Exclusion criteria included the inability to read and understand English, currently listed for a heart transplant, and severe developmental delay (e.g., chart reviewed severe communicative or cognitive dysfunction) that would preclude active participation in the interview. Sample size adequacy was determined when the addition of new data no longer resulted in new categories, insights, or new properties of the core themes, known as "saturation" in some types of qualitative research.<sup>22</sup> Sample size calculation in qualitative research is difficult to predict empirically; the number of interviews required depends on the homogeneity of the population, the purpose of the study, sampling strategy, data quality, types of codes, and goals of saturation (code identification or complete code meaning).<sup>23</sup>

### Data Collection

**Demographic and clinical measures.**—After obtaining informed consent or assent and parental permission, participants were asked to complete a demographic and clinical information form to facilitate the description of the cohort. A medical record review was completed to verify clinical data.

**Qualitative interview.**—Participants were asked to describe examples of how single ventricle heart disease influenced daily school experiences and relationships with others, using an interview guide of open-ended questions developed by the investigators. Additional probing questions were used to clarify and expand their responses. The interview guide was pilot tested during the first four interviews and slightly refined, and then re-submitted to the IRB for approval. All participants received modest monetary compensation for their time, travel, and parking upon completion of the study requirements.

### Data Analysis

The interviews were audio-recorded and were professionally transcribed verbatim. Audio transcripts were analyzed using a method of descriptive phenomenological data analysis

developed by Amedeo Giorgi.<sup>20,24</sup> The steps of data analysis began with assuming the attitude of phenomenological reduction, or true understanding. The data was reviewed in its entirety to get a sense of the whole. Meaningful units within the data were identified with concurrence from the second investigator. The transformations of data into subthemes and then themes were reviewed and discussed with all investigators to ensure that coding was congruent, and differences were resolved through discussion. Finally, the “essence,” or overall meaning of the experience was expressed from the identified themes.<sup>24</sup> The coding of the interview data was completed using NVivo® 12 (QRS International Pty Ltd, Victoria, Australia)<sup>25</sup>, which also provided the audit trail.

Trustworthiness or rigor of qualitative research can be addressed by ensuring credibility, dependability, transferability, confirmability, and reflexivity.<sup>26,27</sup> Credibility, or accuracy, was addressed by having researchers with varied backgrounds participate in data analysis (investigator triangulation) and by going back to participants to verify interpretations (member checks). Transferability is the ability to generalize the findings to the overall population or groups. Dependability is the stability of the study findings over time, and was addressed by having well-defined and described study parameters.<sup>26,27</sup> Confirmability, or ability to be replicated, was enhanced by using an “audit trail” or field notes of the researcher’s observations, ethical concerns and steps taken to manage, data analysis, and how specific pieces of data resulted in specific interpretations.<sup>26,27</sup> Reflexivity, or awareness of how the researcher may influence the findings, was an important consideration in this study because two of researchers have extensive clinical experience caring for children with heart disease. Reflexivity was mitigated using epoché (“to suspend judgment”) and bracketing (putting beliefs aside intentionally), as well as having other members of the study team participate in data analysis.

## Results

### Demographic and Clinical Characteristics

A total of 14 participants completed the study. Table 1 summarizes the demographic and clinical characteristics of the participants. The participants were predominantly white (64%), ranging in age from 14 to 19 years (median 17 years), and all were single and living with parents. The single ventricle diagnoses were highly complex, but 78% were in New York Heart Association functional class 1 or 2. Almost all of the participants (12/14) reported some type of school accommodation, and 28% were either homeschooled or in private school, at least in part due to the impact of single ventricle heart disease and its consequences.

### Interviews

The interviews were 25 to 80 minutes in length. Interview duration varied based on the level of detail in the individual responses. Additional probing questions were asked to clarify detail or elicit additional response if the response was brief or superficial. Although the length of the interviews varied, the themes were consistent across the interviews; the detailed responses of some participants added to the richness of the data collected. The responses of the younger adolescents (14–15 years of age) tended to be more concrete while the older

adolescents' (17–19 years of age) responses were often more detailed and insightful, which was reflected in the longer length of their interviews and added to the depth of the data. After reviewing 11 interview transcripts, no new subthemes were identified, and the data collected appeared to reach saturation. To confirm this, three additional interviews were conducted, which provided additional support for the identified subthemes. From 18 subthemes, four primary themes were identified from the interview transcripts. An overall “essence” was identified and called “optimism despite profound uncertainty.” Table 2 reports subthemes and themes, while Table 3 reports specific interview responses that were associated with each subtheme.

**“Don’t assume”: Pervasive ableism.**—All the participants reported some degree of physical limitation, which varied from fatigue with peak exertion to having difficulty walking in others. Despite these limitations, being labeled as “sick” or “fragile” was a source of frustration, and participants preferred to set their own boundaries. Participants described instances of ableism, where they were not given opportunities or were treated differently because others assumed, they were incapable of performing the proposed task or activity. Several of the adolescents stated that although they knew that they had physical limitations, they wanted to be the one who defined those limitations to others, not to have their limitations decided by others for them.

**“The elephant in the room”: Uncertain future.**—Participants described learning to advocate for their own needs, often from observing their parent’s interactions on their behalf. The adolescents also described a sense of pride and achievement in overcoming physical as well as other types of challenges, often becoming visibly more engaged or smiling while giving these examples. Many also described finding physical activities that were compatible with their endurance limitations such as golf and yoga. Some adolescents described highly protective “helicopter” parents and a struggle to acquire independence. The participants were also quick to identify that their heart disease did not define them, although it was a source of their uniqueness. Future health was a source of uncertainty or anxiety in most, although this generally emerged only after specific questioning, thus being identified as “the elephant in the room.” by one participant. Participants described anxiety in questioning whether fatigue was “real” or whether they could push through it, and if so, what the consequences might be. Some talked about concerns related to whether future health deterioration would affect their plans for careers or living independently. Others talked about perhaps needing a heart transplant, having a shorter lifespan or even early death. Accordingly, responses associated with health uncertainty were accompanied by more intense expressions of sadness or becoming quieter.

**“Everyone finds something to pick on”: Bullying at school.**—Participants described challenges associated with physical education class, in particular, the standardized required task of running a mile. Even with documentation of their activity restriction, some were forced to participate in activities beyond their capability or were not allowed to stop and rest, sometimes resulting in adverse health consequences. A concerning finding was that five of 14 (36%) participants reported being bullied at school, sometimes to the point of needing to change schools. Bullying, generally of the verbal and psychological type, was

reported by both male and female participants, but was more recently experienced by the younger participants. Some of the reasons for bullying included being smaller than other students or not being able to run fast, but one participant related *“everyone finds something to pick on.”* Teachers were often identified as an important source of support, but others were perceived as non-accommodating to legitimate needs. Almost all the participants reported having school accommodations in places, such as having Individualized Education Plan or additional time for tests and assignments. Some participants reported absenteeism due to illness as well as frequent restroom use related to medications. Four participants attended private school or were homeschooled related to the effects of single ventricle heart disease or being unable to succeed in public school. The participants were active in many types of extracurricular and school-based activities typical of adolescents such as band, theater, and Scouts. All participants had goals for the future that included careers (often in health-related professions), living independently, and having their own family. Most identified further education as the major challenge, either academically or financially, to achieving their goals.

**“They know what I have been through”: Social support.**—When asked “Who is the person you talk to when you are having a bad day?” the participants identified parents (5/14, 35.7%), friends (4/14, 28.6%), a teacher (2/14, 14.3%), nobody (2/14, 14.3%), and a therapist (1/14, 7.1%). Parents were often a trusted confidant. The adolescents stated that they often felt most comfortable talking with a parent because of the parent’s familiarity with and shared bond of the experiences of single ventricle heart disease. This reliance upon and importance of parental support continued throughout the age range of participants, who were in different stages of adolescence. Siblings and other family members helped to provide a sense of normalcy. Typical of the adolescent age group, peers were also a crucial source of normalizing daily life and sharing closeness. Several of the participants described a sense of support from their faith and faith-based activities. Another very important source of support was summer heart camp, which many of the participants had attended several times. Participants spoke of support provided by camp staff and felt “safe” because of the medical background of the staff. Many participants spoke about lasting friendships formed with other campers through their common bond of living with heart disease.

**Optimism despite profound uncertainty**—The Figure shows a word cloud created from the participants’ interview quotes related to the four identified themes. The largest words, used most frequently, reflected optimism, like “life,” “can,” “people,” “know,” and “school.” However, there are also some sad or frightening words used quite frequently, like “condition,” “transplant,” and “tired.”

## Discussion

This culturally diverse sample of adolescent Fontan survivors shared detailed experiences related to the impact of single ventricle heart disease on school and social relationships. Our findings concurred with previously reported physical limitations and psychosocial challenges related to living with single ventricle heart disease, and add to the depth of knowledge regarding the impact of these challenges on the daily lives of those with single ventricle heart disease.<sup>5,28–38</sup> New knowledge emerged related to the personal impact of

school challenges such as absenteeism, bullying, being identified as being different or needing accommodations, ableism, and the social benefits of disease-specific cardiac camps that provide opportunities to interact with and establish friendships with like-peers.

### **Ableism/Physical limitations**

Our study findings support previous studies' identification of physical limitations such as varying degrees of exercise endurance and reduced peak performance, as a part of life with single ventricle heart disease survivors.<sup>28,29,32,35-37</sup> Other qualitative studies have described a decreased ability to keep up with peers, which sometimes resulted in social isolation and exclusion.<sup>35,36</sup> In a group of adolescents and adults with single ventricle heart disease, participants reported being able to find and acknowledge their own tolerated activity pace.<sup>29</sup> Participants in the present study also found physical activities, such as golf and yoga, which they could safely engage in and be successful.

Being labeled as "*fragile*" or "*the girl with the heart condition*" and therefore being excluded from social activities or assumptions made by others about one's abilities are types of ableism or discrimination in favor of able-bodied people.<sup>39</sup> Other qualitative studies have reported discrimination or concerns related to employability or insurability in adults with CHD.<sup>32,35</sup> Those who have a chronic illness, such as single ventricle heart disease, should not be assumed to be disabled, or what their disabilities may be.<sup>40</sup>

### **Overcoming obstacles**

Learning to advocate for one's needs is a vital component of developing self-management skills in chronic illness, and to becoming an active and collaborative participant in one's healthcare.<sup>41</sup> Parents, specialty healthcare providers, and school healthcare providers can help facilitate this transition. The adolescents also reported overcoming obstacles to be successful in physical activities appropriate for their abilities, as well as extracurricular activities. These achievements are vital for the development of self-concept and identity, an important developmental task of adolescence.<sup>42</sup>

### **Health uncertainty**

Another important finding of this study, similar to previous qualitative studies, was uncertainty and resulting psychological distress related to future health.<sup>28,29,31-36,38</sup> Concerns over lifelong health challenges or possible early death are common findings in many single ventricle heart disease survivors. Some of the major developmental tasks of adolescence are the search for personal identity, development of self-concept, and becoming independent; chronic illness provides additional challenges to achieving these tasks.<sup>43</sup> The adolescent with single ventricle heart disease may experience conflicts in making plans for their career and independent living due to concerns about their long-term health or even early mortality.

### **School challenges**

A very important finding from the present study, not previously reported in detail, are school-related challenges. One especially problematic area for participants in this study was physical education class, especially the standardized required task of running a mile.



Difficulty in accomplishing this task, or being excluded from it, was often a source of being labeled or identified as different or having difficulty meeting unrealistic expectations. Differences in appearance such as short stature or sternotomy scars, as well as activity limitations or need for school accommodations, made some of the study participants a target for bullying, mostly of the verbal and psychological type. Interestingly, the participants who spoke of bullying did so without much emotionality, which could indicate that it became part of their “normal” school experience, or was “in the past” for older participants. Bullying has been reported in up to 32% of participants in other qualitative studies<sup>33,36</sup>, although the incidence was slightly higher in the present study. Bullying occurs in approximately 20% of school students overall<sup>44</sup>, and the presence of chronic illness or disabilities increases the risk of bullying, although this has not been studied specific to CHD.<sup>13</sup>

Participants in this study reported other impacts of single ventricle heart disease on school success, including difficulty processing information or difficulty focusing on classroom instruction. Most of the participants reported some type of school accommodations such as an Individualized Education Plan, extra time for tests or assignments, or attending special education classes. One previous study reported decreased school quality of life in school-age children with single ventricle heart disease.<sup>45</sup> The need for educational accommodations to assist in academic performance in adolescents with single ventricle heart disease has been reported but needs further investigation on the types of assistance that are most helpful.<sup>5</sup>

### Support systems

Parents were identified as an important source of support across the age range of participants, which stemmed from their intimate familiarity with the experience of growing up with single ventricle heart disease. Friends were also a very important source of social support, as expected for the adolescent age group; close friendships improved their sense of belonging. Other studies have reported the vital role of family and friends in providing a social support network for adolescents and adults with single ventricle heart disease and other complex CHD.<sup>29,32,34,35,38</sup> A study that examined “clusters” of social support in adolescents with CHD identified that, compared to healthy controls, adolescents with CHD were more likely to have a cluster of combined parental and peer support as opposed to primarily only parental support, peer support, or lack of support.<sup>46</sup> This study also reported that adolescents in the combined support cluster had lower depression symptoms and higher reported well-being.<sup>46</sup>

Many of the study participants had previously attended a cardiac-specific, medically supervised summer camp sponsored by a non-profit organization. Potential benefits of these disease-specific camps include improved self-esteem, self-concept, and quality of life by providing safe opportunities to succeed in activities they may not have access to otherwise, such as climbing walls and zip-lining, developing social relationship skills with peers like themselves, and improving disease knowledge and management skills.<sup>47–49</sup> The beneficial effects of this type of peer support should not be underestimated; one of the most stoic participants in this study became visibly tearful when talking about the importance of heart camp and the friendships she formed there.

## The influence of adolescence on participants

The participants ranged in age from 14 to 19 years, which spans the developmental period of adolescence; yet there were some differences between the participant's responses at different ends of this age spectrum. The younger participants, usually in the 8<sup>th</sup> or 9<sup>th</sup> grade, tended to be more concrete in their answers but also had more recently experienced challenges such as participating in physical education class and being bullied. Bullying behaviors tend to peak in middle school and decrease throughout high school, so the responses of the younger participants may reflect these age-related differences.<sup>50</sup> The older participants (17–19 years old) tended to be more reflective and insightful, and this was evident in the length and depth of their interview responses. The period of adolescence includes progressive changes in cognitive abilities, such as the ability to think hypothetically and abstractly, and to develop formal operational thinking.<sup>51</sup> Psychosocial development during adolescence includes establishing autonomy and a shift in importance of family and peers, development of a sense of identity including self-esteem and self-concept, as well as development of emotional self-regulation and ability to form effective social relationships.<sup>51</sup> These developmental tasks may also be adversely affected or delayed in the setting of CHD and its associated psychological and cognitive comorbidities. None of the participants had yet attained fully independent living, so they were all classified as adolescents despite some variance in their developmental level.

## Implications of study findings

Although the adolescent with single ventricle heart disease may have subnormal exercise tolerance, promotion of physical activity appropriate to the individual's health status, or an activity prescription as opposed to a restriction, is beneficial for cardiac health as well as overall well-being.<sup>52</sup> Cognitive screening to identify evolving or new neurodevelopmental sequelae of single ventricle heart disease can be done quickly during outpatient visits and appropriate referrals made.<sup>53</sup> A model successfully implemented in several cardiac neurodevelopmental follow-up programs is the incorporation of the school liaison role (e.g. typically a teacher) to work directly with schools to communicate educational recommendations from neurodevelopmental testing, act as a family advocate, and be a liaison between the hospital, family and the school.<sup>54</sup> School nurses can provide support to enacting school accommodations in place without resulting in social isolation, helping teachers understand the student's physical limitations, and developing appropriate emergency medical care plans.

Healthcare providers can assist adolescents with single ventricle heart disease to be active participants during healthcare visits and advocate for the adolescent's autonomy with parents or caregivers.<sup>5</sup> Anxiety and depression should be anticipated, screened for, discussed, and appropriate referrals made.<sup>5</sup> The adolescent with single ventricle heart disease should be provided with accurate information about their current and anticipated future health. Heart camp referrals and attendance can facilitate social engagement with other adolescents like themselves.

The American Heart Association published best practices for transition of adolescents and young adults with CHD to adult cardiac care.<sup>55</sup> Transition programs should consider the

need for psychological support and the beneficial influence of peers, a peer-led support group with the input of a multi-disciplinary team might be helpful. Some heart camps also offer a transition program to assist young adults with transition-related planning, such as obtaining health insurance, employability, and living skills. Given the variation in developmental achievement and the potential effects of developmental co-morbidities, it is important to consider the adolescent's unique developmental level, and to tailor a transition program that meets their individual needs. It is also important to determine, with the adolescent's input, what level of parental involvement is appropriate at different phases of transition planning.

The unique school-related challenges that emerged from this study, including physical challenges and health, absenteeism, bullying, being identified as different, and ableism, warrant additional investigation. Adding the perspectives of teachers, school nurses, and parents would provide valuable information. Strategies or programs that may support success in school and college should be evaluated. In addition, transition programs from pediatric to adult cardiac care must be evaluated, and the effectiveness of strategies to improve long-term follow-up should be studied.

### Limitations

The adolescents who participated in the study were not representative of all Fontan survivors; most participants had previously participated in research and were known to be able to describe their experiences. The participants likely over-represent Fontan survivors who were medically stable within their chronic illness. The length and depth of responses may reflect the participant's response to the novel situation of being interviewed by a previously unknown person, as well as other factors such as level of fatigue or energy, willingness to disclose personal information, or levels of personal insight. The novel situation may have also influenced the participant's willingness to demonstrate or share emotionality, since the interviewer was not previously known to the participant.

Phenomenological research requires interpretation, which may lead to researcher bias. However, non-cardiac members of the investigative team concurred on the common subthemes and themes that emerged. Phenomenological research is not generalizable but does yield valuable understanding into lived experiences and may help guide theory development and future research questions.

### Conclusion

Adolescents and young adults with single ventricle heart disease express optimism despite uncertainties related to school and social relationships as well as their future health. These adolescents face many challenges as well as an uncertain future, yet they can adapt to their activity limitations and find ways to succeed in the context of their abilities. However, some of them also experience anxiety and sadness related to uncertainty about their future. Support systems, in particular family, peers, and teachers, provide them with resources to cope with the challenges associated with single ventricle heart disease. These findings support previous qualitative research in participants with single ventricle heart disease and

add new information on experiences of school and social relationships not previously reported.

## Acknowledgments:

The authors gratefully acknowledge the assistance of Anna Strömberg, Ph.D., RN, Professor, Division of Nursing, Linköping University, Linköping, Sweden, for her assistance in reviewing and coding some of the interview transcripts. In addition, Paola Moreno, BA, University of California, Los Angeles, Staff Research Assistant, provided valuable support in recruiting subjects and translation. The authors would also like to thank the study participants and their families for contributing their time and effort in the interviews.

### Financial Support:

This work was partially supported by National Institutes of Health R01NR013930 and R01NR016463 (NAP), American Heart Association Pre-Doctoral fellowship 15PRE25300024 (JKP), Jonas Nurse Leader Scholarship, Cohort IV (JKP), and University of California Irvine Public Impact Fellowship 2017–2018 (JKP)

## References

- Schilling C, Dalziel K, Nunn R, et al. The Fontan epidemic: Population projections from the Australia and New Zealand Fontan Registry. *Int J Cardiol.* 2016;219:14–19. doi:10.1016/j.ijcard.2016.05.035 [PubMed: 27257850]
- Pundi KN, Johnson JN, Dearani JA, et al. 40-year follow-up after the Fontan operation: Long-term outcomes of 1,052 patients. *J Am Coll Cardiol.* 2015;66(15):1700–1710. doi:10.1016/j.jacc.2015.07.065 [PubMed: 26449141]
- Gewillig M, Goldberg DJ. Failure of the Fontan circulation. *Heart Fail Clin.* 2014;10(1):105–116. doi:10.1016/j.hfc.2013.09.010 [PubMed: 24275298]
- Kreutzer C, Kreutzer G. The lymphatic system: The Achilles Heel of the Fontan-Kreutzer circulation. *World J Pediatr Congenit Heart Surg.* 2017;8(5):613–623. doi:10.1177/2150135117720685 [PubMed: 28901223]
- Rychik J, Atz AM, Celermajer DS, et al. Evaluation and management of the child and adult with Fontan circulation: A Scientific Statement from the American Heart Association. *Circulation.* 2019;140(6). doi:10.1161/CIR.0000000000000696
- Marino BS, Lipkin PH, Newburger JW, et al. Neurodevelopmental outcomes in children with congenital heart disease: Evaluation and management a Scientific Statement From the American Heart Association. *Circulation.* 2012;126(9):1143–1172. doi:10.1161/CIR.0b013e318265ee8a [PubMed: 22851541]
- Kharitonova M, Marino BS. An emergent phenotype: A critical review of neurodevelopmental outcomes for complex congenital heart disease survivors during infancy, childhood, and adolescence In: *Congenital Heart Disease and Neurodevelopment.* Elsevier; 2016:55–87. doi:10.1016/B978-0-12-801640-4.00005-6
- Pike NA, Roy B, Gupta R, et al. Brain abnormalities in cognition, anxiety, and depression regulatory regions in adolescents with single ventricle heart disease. *J Neurosci Res.* 2018;96(6):1104–1118. doi:10.1002/jnr.24215 [PubMed: 29315714]
- DeMaso DR, Calderon J, Taylor GA, et al. Psychiatric disorders in adolescents with single ventricle congenital heart disease. *Pediatrics.* 2017;139(3). doi:10.1542/peds.2016-2241
- Hövels-Gürich HH, McCusker C. Neurodevelopmental patterns in congenital heart disease across childhood: Longitudinal studies from Europe In: McCusker C, Casey F, eds. *Congenital Heart Disease and Neurodevelopment.* Academic Press; 2016:41–53. doi:10.1016/B978-0-12-801640-4.00004-4
- Bellinger DC, Watson CG, Rivkin MJ, et al. Neuropsychological status and structural brain imaging in adolescents with single ventricle who underwent the Fontan procedure. *J Am Heart Assoc Cardiovasc Cerebrovasc Dis.* 2015;4(12). doi:10.1161/JAHA.115.002302
- Mahle WT, Clancy RR, Moss EM, Gerdes M, Jobs DR, Wernovsky G. Neurodevelopmental outcome and lifestyle assessment in school-aged and adolescent children with hypoplastic left

- heart syndrome. *Pediatrics*. 2000;105(5):1082–1089. doi:10.1542/peds.105.5.1082 [PubMed: 10790466]
13. Pittet I, Berchtold A, Akre C, Michaud P-A, Suris J-C. Are adolescents with chronic conditions particularly at risk for bullying? *Arch Dis Child*. 2010;95(9):711–716. doi:10.1136/adc.2008.146571 [PubMed: 19307194]
  14. Bellinger DC. Are children with congenital cardiac malformations at increased risk of deficits in social cognition? *Cardiol Young*. 2008;18(1):3–9. doi:10.1017/S104795110700176X [PubMed: 18093362]
  15. Maslow GR, Haydon A, McRee A-L, Ford CA, Halpern CT. Growing up with a chronic illness: social success, educational/vocational distress. *J Adolesc Health Off Publ Soc Adolesc Med*. 2011;49(2):206–212. doi:10.1016/j.jadohealth.2010.12.001
  16. Maslow GR, Haydon A, Ford CA, Halpern CT. Young adult outcomes of children growing up with chronic illness: An analysis of the National Longitudinal Study of Adolescent Health. *Arch Pediatr Adolesc Med*. 2011;165(3):256–261. doi:10.1001/archpediatrics.2010.287 [PubMed: 21383274]
  17. Husserl E *Logical Investigations*. Vol 1 Humanities Press; 1970.
  18. Christensen M, Welch A, Barr J. Husserlian descriptive phenomenology: A review of intentionality, reduction and the natural attitude. *J Nurs Educ Pract*. 2017;7(8):113. doi:10.5430/jnep.v7n8p113
  19. Creswell JW, Poeh C. *Qualitative Inquiry and Research Design: Choosing Among Five Approaches*. 4th Edition SAGE Publications, Inc; 2017.
  20. Christensen M The empirical-phenomenological research framework: Reflecting on its use. *J Nurs Educ Pract*. 2017;7(12):81. doi:10.5430/jnep.v7n12p81
  21. Cabrera-Mino C, Roy B, Woo MA, et al. Reduced brain mammillary body volumes and memory deficits in adolescents who have undergone the Fontan procedure. *Pediatr Res*. Published online 9 9, 2019. doi:10.1038/s41390-019-0569-3
  22. Morse JM. The significance of saturation. *Qual Health Res*. 1995;5(2):147–149. doi:10.1177/104973239500500201
  23. Hennink MM, Kaiser BN, Marconi VC. Code saturation versus meaning saturation: How many interviews are enough? *Qual Health Res*. Published online 9 25, 2016. doi:10.1177/1049732316665344
  24. Giorgi A *The Descriptive Phenomenological Method in Psychology: A Modified Husserlian Approach*. Duquesne University Press; 2009.
  25. NVivo qualitative data analysis software | QSR International. Accessed June 6, 2019 <https://www.qsrinternational.com/nvivo/home>
  26. Guba EG, Lincoln YS. Epistemological and methodological bases of naturalistic inquiry. *ECTJ*. 1982;30(4):233–252. doi:10.1007/BF02765185
  27. Cypress BS. Rigor or reliability and validity in qualitative research: Perspectives, strategies, reconceptualization, and recommendations. *Dimens Crit Care Nurs*. 2017;36(4):253–263. doi:10.1097/DCC.0000000000000253 [PubMed: 28570380]
  28. Berghammer MC, Brink E, Rydberg AM, Dellborg M, Ekman I. Committed to life: Adolescents' and young adults' experiences of living with Fontan circulation. *Congenit Heart Dis*. 2015;10(5):403–412. doi:10.1111/chd.12244 [PubMed: 25564938]
  29. Overgaard D, King C, Christensen RF, Schrader A-M, Adamsen L. Living with half a heart--experiences of young adults with single ventricle physiology: a qualitative study. *J Cardiovasc Nurs*. 2013;28(2):187–196. doi:10.1097/JCN.0b013e3182498677 [PubMed: 22635059]
  30. Pike NA, Evangelista LS, Doering LV, Eastwood J-A, Lewis AB, Child JS. Quality of life, health status, and depression: comparison between adolescents and adults after the Fontan procedure with healthy counterparts. *J Cardiovasc Nurs*. 2012;27(6):539–546. doi:10.1097/JCN.0b013e31822ce5f6 [PubMed: 21912272]
  31. du Plessis K, Peters R, King I, et al. "How long will I continue to be normal?" Adults with a Fontan circulation's greatest concerns. *Int J Cardiol*. 2018;260:54–59. doi:10.1016/j.ijcard.2018.01.098 [PubMed: 29622455]

32. Claessens P, Moons P, de Casterlé BD, Cannaerts N, Budts W, Gewillig M. What does it mean to live with a congenital heart disease? A qualitative study on the lived experiences of adult patients. *Eur J Cardiovasc Nurs*. 2005;4(1):3–10. doi:10.1016/j.ejcnurse.2004.12.003 [PubMed: 15718186]
33. Chiang YT, Chen CW, Su WJ, et al. Between invisible defects and visible impact: the life experiences of adolescents and young adults with congenital heart disease. *J Adv Nurs*. 2015;71(3):599–608. doi:10.1111/jan.12546 [PubMed: 25296699]
34. Cornett L, Simms J. At the ‘heart’ of the matter: An exploration of the psychological impact of living with congenital heart disease in adulthood. *J Health Psychol*. 2014;19(3):393–406. doi:10.1177/1359105312471569 [PubMed: 23407131]
35. Lee S, Kim S-S. The life experiences of Korean children and adolescents with complex congenital heart disease: A qualitative study: Life experiences of patients. *Nurs Health Sci*. 2012;14(3):398–404. doi:10.1111/j.1442-2018.2012.00709.x [PubMed: 22950619]
36. McMurray R, Kendall L, Parsons JM, et al. A life less ordinary: growing up and coping with congenital heart disease. *Coron Health Care*. 2001;5(1):51–57. doi:10.1054/chec.2001.0112
37. Shearer K, Rempel GR, Norris CM, Magill-Evans J. “It’s no big deal”: Adolescents with congenital heart disease. *J Pediatr Nurs*. 2013;28(1):28–36. doi:10.1016/j.pedn.2012.03.031 [PubMed: 22543260]
38. Zahmacioglu O, Yildiz CE, Koca B, et al. Coming from behind to win—a qualitative research about psychological conditions of adolescents who have undergone open-heart surgery for single ventricle between the ages 0–5. *J Cardiothorac Surg*. 2011;6:155. doi:10.1186/1749-8090-6-155 [PubMed: 22112589]
39. Friedman C, Owen AL. Defining disability: Understandings of and attitudes towards ableism and disability. *Disabil Stud Q*. 2017;37(1). doi:10.18061/dsq.v37i1.5061
40. Wendell S. Unhealthy disabled: Treating chronic illnesses as disabilities. *Hypatia*. 2001;16(4):17–33. doi:10.1111/j.1527-2001.2001.tb00751.x
41. McCabe EM. School nurses’ role in self-management, anticipatory guidance, and advocacy for students with chronic illness. *NASN Sch Nurse*. Published online 2 18, 2020:1942602X2090653. doi:10.1177/1942602X20906534
42. Holmes BW, Duncan PM. Promoting the health of adolescents In: McInerney TK, Adam HM, Campbell D, DeWitt TG, Foy JM, Kamat D, eds. *American Academy of Pediatrics Textbook of Pediatric Care*. 2nd ed American Academy of Pediatrics; 2017:187–191.
43. Balk DE. *Dealing with Dying, Death, and Grief during Adolescence*. 1st Edition Routledge; 2014.
44. *Bullying Statistics*. Published 2017. Accessed April 20, 2019 <https://www.pacer.org/bullying/resources/stats.asp>
45. Gerstle M, Beebe DW, Drotar D, Cassidy A, Marino BS. Executive functioning and school performance among pediatric survivors of complex congenital heart disease. *J Pediatr*. 2016;173:154–159. doi:10.1016/j.jpeds.2016.01.028 [PubMed: 26875011]
46. Oris L, Seiffge-Krenke I, Moons P, et al. Parental and peer support in adolescents with a chronic condition: a typological approach and developmental implications. *J Behav Med*. 2016;39(1):107–119. doi:10.1007/s10865-015-9680-z [PubMed: 26369633]
47. McCarthy A. Summer camp for children and adolescents with chronic conditions. *Pediatr Nurs*. 2015;41(5):245–250. [PubMed: 26665424]
48. Desai PP, Sutton LJ, Staley MD, Hannon DW. A qualitative study exploring the psychosocial value of weekend camping experiences for children and adolescents with complex heart defects. *Child Care Health Dev*. 2014;40(4):553–561. doi:10.1111/cch.12056 [PubMed: 23551299]
49. Bultas MW, Budhathoki C, Balakas K. Evaluation of child and parent outcomes after a pediatric cardiac camp experience. *J Spec Pediatr Nurs*. 2013;18(4):320–328. doi:10.1111/jspn.12040 [PubMed: 24094127]
50. Menesini E, Salmivalli C. Bullying in schools: the state of knowledge and effective interventions. *Psychol Health Med*. 2017;22(sup1):240–253. doi:10.1080/13548506.2017.1279740 [PubMed: 28114811]
51. Sanders RA. Adolescent psychosocial, social, and cognitive development. *Pediatr Rev*. 2013;34(8):354–359. doi:10.1542/pir.34-8-354 [PubMed: 23908362]

52. Longmuir PE, Brothers JA, de Ferranti SD, et al. Promotion of physical activity for children and adults with congenital heart disease. *Circulation*. 2013;127(21):2147–2159. doi:10.1161/CIR.0b013e318293688f [PubMed: 23630128]
53. Pike NA, Poulsen MK, Woo MA. Validity of the Montreal Cognitive Assessment screener in adolescents and young adults with and without congenital heart disease. *Nurs Res*. 2017;66(3):222–230. doi:10.1097/NNR.000000000000192 [PubMed: 28448372]
54. School Intervention Program | Children’s Hospital of Wisconsin. Published 2019. Accessed April 27, 2019 <https://chw.org/medical-care/herma-heart/programs/school-intervention-program>
55. Sable C, Foster E, Uzark K, et al. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. *Circulation*. 2011;123(13):1454–1485. doi:10.1161/CIR.0b013e3182107c56 [PubMed: 21357825]



**Figure.**  
 Word cloud generated from interview quotations of adolescents with single ventricle heart disease that reflect the themes identified. The size of the word increases with the frequency that the word appears.



**Table 1.**

Demographic and Clinical Characteristics of Study Participants (N = 14)

Characteristic	Results
<b>Age</b> (years) median, (IQR)	17 (15, 18)
<b>Male gender</b> N, (%)	7 (50.0)
<b>Race/Ethnicity</b> N, (%)	
White	9 (64.3)
Hispanic	3 (21.4)
Black	2 (14.3)
<b>Cardiac diagnosis</b> N, (%)	
Hypoplastic left heart syndrome	4 (28.6)
Unbalanced AVSD, heterotaxy	3 (21.4)
Double outlet right ventricle	3 (21.4)
Double inlet left ventricle	2 (14.3)
Other complex	2 (14.3)
<b>Health insurance type</b> N, (%)	
Public	6 (42.9)
Private	7 (50.0)
Unsure	1 (7.1)
<b>NYHA functional class</b>	
1 No limitations of physical activity	1 (7.1)
2 Mild limitations of physical activity	10 (71.5)
3 Marked limitations of physical activity	3 (21.4)
4 Symptoms at rest	0
<b>School accommodations</b> <sup>1</sup> N (%)	12 (85.7)
Individualized Education Plan	6 (42.9)
Extra time for assignments	6 (42.9)
Extra time for tests	7 (50.0)
Special Education classes	3 (21.4)
Therapy at school (PT/OT/speech)	1 (7.1)
No physical education class	3 (21.4)
Teaching Assistant sits with student	1 (7.1)
<b>Type of school</b> N (%)	
Public school	10 (71.4)
Home schooled	2 (14.3)
Private school	2 (14.3)
<b>Activity restriction</b> N, (%)	
Self-limit	9 (64.3)
Specific prescribed restriction	3 (21.4)
No restriction	2 (14.3)

Given are frequency (N) and percent (%) for categorical variables, median and interquartile range (IQR) for continuous variables. AVSD = atrioventricular septal defect; NYHA = New York Heart Association; PT = Physical therapy; OT = Occupational therapy.

<sup>1</sup> participants may have more than one type of accommodation.

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

**Table 2.**

Optimism despite profound uncertainty themes and subthemes

Theme	Subthemes
1. "Don't assume": Pervasive ableism	Physical limitations Labeled as sick Difference from peers
2. "The elephant in the room": Uncertain future	Self-advocacy Despite some limitations, achieving my goals Overprotective parents Single ventricle heart disease doesn't define me Concerns for future health
3. "Everyone finds something to pick on": Bullying at school	Physical education class Being bullied or picked on Teachers: facilitate accommodations or create barriers Absenteeism and other obstacles Extracurricular activities Plans for future
4. "They know what I have been through": Social support	Parents and family relationships Peer relationships Faith Heart camp: Friends like me

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 3.

## Themes, subthemes, and supporting quotations

Theme	Subthemes and supporting quotations
1. "Don't assume": Pervasive ableism	<p><b>Physical limitations:</b> "I do get tired more easily, but I can do pretty much anything, honestly." "I can go a million miles, but only for a short time."</p> <p><b>Being labeled as sick:</b> "She's the girl with the heart condition" "... You're too delicate, don't worry, I'll do it because you can't..."</p> <p><b>Being different from peers:</b> "I typically don't like people to know that I'm on accommodations."</p>
2. "The elephant in the room": Uncertain future	<p><b>Advocating for my needs:</b> "I really kind of ask for everything on my own"</p> <p><b>Despite some limitations, achieving my goals:</b> "But I stayed persistent at it [golf] and kept trying because it's something I wanted to do, and it was an outlet that I could do."</p> <p><b>Overprotection:</b> "They have that 'helicopter' parenting style, specifically with me... they're always watching to see if I'm tired."</p> <p><b>SVHD doesn't define me:</b> "I never thought of it as a weird thing that made me completely different. It was more of "Yeah, I have heart problems; it's a thing."</p> <p><b>Concerns for future health:</b> "Well, they have told me that I'll probably have to have a heart transplant, and if I don't, I'll probably die sooner. That's scary to think about... I've cried sometimes..." "I was doing research on my own heart condition. And I learned the definition of 'palliative'... And I read a thing that said, 'All Fontans fail at some point.' But that kind of stuff, I wasn't ready to see that."</p>
3. "Everyone finds something to pick on": Bullying at school	<p><b>Physical education class:</b> "... when I was younger, I had to be in PE, and one time someone didn't treat me different-they told me to keep going, and I passed out."</p> <p><b>Being bullied or picked on:</b> "I would hear them whispering, 'Hey guys, when we go, run really fast, so X doesn't get a seat at the library.' Because they knew that I needed to sit down." "... I was getting bullied from these kids, they would see me as the weak one... [my Mom] would see me crying at home... We tried moving schools. That helped a bit."</p> <p><b>Teachers: facilitate accommodations or create barriers:</b> "They [teachers] would like to think that I was using my heart condition as an excuse to not do things." "She [a helpful teacher] moves a lot of stuff [assignment due dates] around for me."</p> <p><b>Absenteeism and other school obstacles:</b> "Sometimes I have a hard time retaining information..." "I would miss a lot of days because I have to go to the emergency room a lot..."</p> <p><b>Extracurricular activities:</b> "I'm one of the editors of my [school] newspaper." "I have band practice... I'm mainly in percussion. But right now, they have me playing the piano." "...one of the teachers started a yoga class, and so I took that up."</p> <p><b>Plans for the future:</b> "I'd like to be in the medical field in some way." "The biggest challenge for the future?... probably the whole getting through college thing."</p>
4. "They know what I've been through": Social support	<p><b>Parents and family relationships:</b> "I feel like I can talk to both of my parents, my Mom, and my Dad... They're understanding... Because they know what I've been through." "We treat each other like brothers normally do by beating the living hell out of each other."</p> <p><b>Peers:</b> "[my friends] treat me like everyone else. We laugh. We do really dumb stuff together." "People, when they say, 'I love you,' and they do this [makes heart symbol with two curved hands together]. My friends do this to me [makes half the heart symbol with one hand] because there's half. So, we do that..."</p> <p><b>Faith:</b> "I've always just really believed that I'm going to be OK. God's got me."</p> <p><b>Heart camp: Friends like me:</b> "Camp introduced me to people like me, as well as growing my confidence." "[heart camp] taught me not only what my condition was, but how to live with it."</p>