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IRVINE

Thriving in the Face of Uncertainty: Life Experiences of Adolescents with Single Ventricle Heart Disease

DISSERTATION

submitted in partial satisfaction of the requirements
for the degree of

DOCTOR OF PHILOSOPHY

in Nursing Science

by

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DEDICATION

To

my parents, Rich and Anne Peterson

for instilling and nurturing my belief that I can accomplish

whatever I desire through hard work and determination.

To my family, friends and colleagues who supported me on this journey in so many ways.

There are not enough words to thank all of you.

And to the children and their families affected by congenital heart disease

that I have had the privilege to work with in my career,

especially those who participated in this research.

Their amazing strength and resilience through unbelievably

difficult times has always been an inspiration for me.

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PUBLICATIONS

Journal articles:

Peterson J, Adlard K, Walti BI, Hayakawa J, McClean E, Feidner SC. Clinical nurse specialist collaboration to recognize, prevent, and treat pediatric pressure ulcers. *Clinical Nurse Specialist*. 2015; 29(5): 276-282. doi 10.1097/NUR.0000000000000135

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ABSTRACT OF THE DISSERTATION

Thriving in the Face of Uncertainty: Life Experiences of Adolescents with Single Ventricle Heart Disease

By

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Doctor of Philosophy in Nursing Science

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Professor Lorraine S. Evangelista, Chair

Background: Survivors of single ventricle heart disease (SVHD) must cope with challenges related to physical health, neurodevelopmental delays, and psychosocial sequelae of SVHD, which also affect academic achievement and social relationships. Despite these challenges, SVHD survivors often rate their health and quality of life (QOL) relatively highly. However, there are few studies that examine the life experiences of adolescents with SVHD.

Purpose: The purpose of this study was to examine the lived experiences of school, social relationships and healthcare in adolescents with SVHD.

Methods: A descriptive phenomenological methodology was employed, utilizing interviews of adolescents who had previously undergone a Fontan procedure for SVHD. The interviews were analyzed using the phenomenological analysis technique developed by Giorgi. Participants also completed a demographic and clinical information form that included one-dimensional questions related to overall health, changes in health, and overall QOL. A medical chart review was completed to obtain information on medical and surgical history and current status.

Results: Fourteen adolescents (ages 14 to 19 years, seven males) participated. The interviews lasted from 24 to 81 minutes. Self-reported health was rated as excellent to good in 12/14 participants and median self-reported QOL was 87.5 on a 0 to 100 scale. Seven themes emerged from the interviews, including 1) I am learning to take care of myself; 2) I have some limitations, but don't assume: dealing with ableism; 3) I see myself as healthy; 4) I have big plans for the future, but there is also health uncertainty; 5) School can be a challenge; 6) My QOL is very good, and: 7) I have lots of healthcare experiences. The overall essence from these themes was "Thriving in the Face of Uncertainty."

Conclusions: Adolescents with SVHD identified their physical limitations, school challenges, and the need to learn self-care in the face of an uncertain future related to their health. Despite these physical and psychosocial limitations, they consider themselves healthy with a high QOL and remain optimistic for the future, drawing strength from family, friend, teachers, and healthcare providers for support.

Chapter 1. Introduction

Single ventricle heart disease (SVHD) is a broad term for complex congenital heart disease (CHD) in which complete mixing of systemic and pulmonary venous return occurs due to hypoplasia and/or atresia of one ventricle and/or atrioventricular valve (Schwartz, McCracken, Petit, & Sachdeva, 2017). Surgical palliation of SVHD usually involves three complex operations in infancy and early childhood, culminating in the total cavo-pulmonary connection, or Fontan procedure. The Fontan procedure separates the venous and arterial circulations by redirecting pulmonary blood flow via passive drainage and allowing the single ventricle to pump blood systemically.

Survival in CHD has improved dramatically over the past 30 years, due to improvements in recognition, surgical techniques, and perioperative management. Improved survival has resulted in a rapid increase in the prevalence of adolescents and young adults with CHD, including an increasing percentage of CHD survivors with more complex forms of CHD including SVHD (Marino et al., 2012; Sadowski, 2009; Wu, Lu, Chen, Kao, & Huang, 2018). Likewise, long-term survival for patients with SVHD has improved greatly over time, as advances in medical and surgical care have evolved. The average 15-year survival following the Fontan procedure has been reported at 84.1% (Schwartz et al., 2017), and another meta-analysis reported 20-year survival at 82% (Poh & d'Udekem, 2017). Although mortality has improved from the 1980s, children with SVHD born in the 2000s are still 30 times more likely to die than age- and gender-matched general United States population controls (Spector et al., 2018).

Children with SVHD are at the highest risk for adverse neurodevelopmental outcomes because of the complexity of their medical treatment and the need for open heart surgery in the neonatal or early infancy time period; it is estimated that only a minority of patients with SVHD

are completely free of neurodevelopmental delays (Marino et al., 2012). The American Academy of Pediatrics and the American Heart Association have developed guidelines for neurodevelopmental assessment and referral for intervention in all children with CHD, to mitigate the functional impact of neurodevelopmental delays associated with CHD and its treatment. These guidelines stress the importance of longitudinal assessment from infancy to adulthood, because previously identified problems can evolve and new problems can emerge as developmental and functional demands increase (Hövels-Gürich & McCusker, 2016; Marino et al., 2012) However, implementation of these guidelines has been challenging (Knutson, Kelleman, & Kochilas, 2016), and the current generation of SVHD survivors are too old to have benefited from these recent guidelines. Fontan survivors must cope with additional complications of their SVHD and its treatment, including arrhythmias, protein-losing enteropathy, and other lymphatic disorders, heart failure, plastic bronchitis, thromboembolism and hepatic cirrhosis (Anderson et al., 2008; Driscoll, 2007; O'Leary, 2002; Pike et al., 2011). These sequelae lead to reduced exercise capacity as well as reduced functional status, which in turn, affect the ability to participate in activities, school, and employment (Atz et al., 2015; Driscoll, 2007; Williams et al., 2009).

Adolescents with SVHD also face challenges in the academic setting. These challenges include a variety of cognitive deficits such as attention, executive functioning, language, memory, and conceptual thinking (Kharitonova & Marino, 2016; Marino et al., 2012). Adolescents with SVHD were found to have significantly lower scores on tests of reading and mathematics, compared to age matched normal controls (Bellinger et al., 2015). Two recent studies identified that adolescents with SVHD had significantly lower scores than age-matched normal controls in most areas of cognitive function and memory (Pike et al., 2018; Singh et al.,

2018) Another recent study reported that, compared to age-matched normal controls, adolescents with SVHD have higher lifetime and current rates of anxiety disorders (35% vs. 7% lifetime, 22% vs. 6% current) and attention deficit hyperactivity disorder (ADHD) (34% vs. 6% lifetime, 33% vs. 4% current). In addition, adolescents with SVHD reported more anxiety and depressive symptoms and more disruptive behaviors than age-matched normal controls (DeMaso et al., 2017). These neurodevelopmental, cognitive, and psychological challenges impact all areas of development and daily living. Because the "jobs" of adolescents are school and school-related activities, the challenges associated with SVHD also impact scholastics success as well as the development of social relationships with family, teachers, and peers.

Some studies have identified that children or adolescents with physical disabilities or chronic medical conditions are at increased risk for being bullied, due to differences in physical appearance and abilities as well as impaired cognitive or social functioning (Faith et al., 2015; Pittet, Berchtold, Akre, Michaud, & Suris, 2010; Sentenac et al., 2011). Bullying is "aggressive behavior or intentional harm doing by peers that is carried out repeatedly and involves an imbalance of power, either actual or perceived, between the victim and the bully," carried out to establish a social network or hierarchy (Olweus, 1993). Adolescents with SVHD have many risk factors for being identified as "different" including usually small stature, decreased exercise capacity, exclusion from some physical activities such as physical education class, or need for special education services, and may thus be a target for bullying.

In large part due to improved survival and recognition of neurodevelopmental sequelae of CHD and its treatment, especially for adolescents with SVHD, quality of life (QOL) has become an important research focus. Variations in definitions of QOL versus health-related QOL (HRQOL) versus perceived health have clouded many investigations. The few studies examining

QOL in SVHD survivors had methodological challenges and small sample sizes. Likewise, studies of QOL and HRQOL have included a spectrum of CHD severity which limits generalizability to the SVHD population. Congenital heart disease survivors and Fontan survivors often rated their overall QOL as not significantly different from healthy controls, despite the fact that they rated themselves lower in physical functioning and proxies and healthcare providers rated their QOL lower (Apers et al., 2016; Moons et al., 2006; Pike et al., 2012). However, other studies also reported lower perceived physical health and HRQOL in patients with complex CHD including SVHD (Bruto, Harrison, Fedak, Rockert, & Siu, 2007; Hager & Hess, 2005; Jackson, Hassen, Gerardo, Vannatta, & Daniels, 2016; Mellion et al., 2014; Müller, Berner, Ewert, & Hager, 2014).

Purpose of the Study

Because of the ongoing medical and psychosocial challenges that SVHD survivors face, their experiences of school, relationships and healthcare may be very different from the same experiences in people without SVHD. Little is known about the lived experiences of this first generation of SVHD survivors; existing qualitative studies include participants from outside the United States, or include the spectrum of CHD severity (Berghammer, Brink, Rydberg, Dellborg, & Ekman, 2015; Chiang et al., 2015; Claessens et al., 2005; Cornett & Simms, 2014; du Plessis et al., 2018; Lee & Kim, 2012; McMurray et al., 2001; Overgaard, King, Christensen, Schrader, & Adamsen, 2013; Shearer, Rempel, Norris, & Magill-Evans, 2013; Zahmacioglu et al., 2011).

The proposed study will examine the lived experiences of Fontan survivors to gain a better understanding of how SVHD affects their experiences of school, relationships, and interactions with the healthcare system using a phenomenological methodology. The unique medical physiology of Fontan survivors requires a lifetime of medical care and procedures. Given the

long-term physical and psychological health effects of SVHD, it is vital for healthcare providers to be aware of and understand the unique life experiences of Fontan survivors to provide comprehensive and integrative care and to target interventions to help them overcome obstacles and achieve their maximal functional outcomes.

The **specific aims of this study** are to:

1. Assess the perceptions of daily life of adolescents with SVHD.
2. Examine the influence of SVHD on daily life experiences including school and social relationships in adolescents with SVHD.
3. Explore the influence of SVHD on healthcare experiences of adolescents with SVHD.

The **research questions** for this study are:

1. What are the lived experiences of adolescents with SVHD?
2. What are the successes and challenges associated with school and social relationships in adolescents with SVHD?
3. How do adolescents with SVHD perceive their health and QOL?
4. How do adolescents with SVHD perceive their healthcare experiences?

Significance of the Study for Practice and Research

Despite their growing numbers, there is very little research on how survivors of SVHD perceive their experiences in school, relationships with others and healthcare encounters. These adolescents are the first generation with such complex CHD to reliably survive to adulthood, and they face an uncertain future. The few studies that have reported on lived experiences of adolescents with SVHD were undertaken by researchers outside the United States, and differences in social support mechanisms and healthcare systems between Europe and the United States limit the generalizability of the European findings. This study will add the perspective of

adolescents living in the United States to the body of knowledge. In addition, this study will provide needed information that will assist healthcare and service providers to understand, anticipate and meet the needs of this unique population to allow them to achieve their maximum health potential and contribute to society at the highest level possible.

Chapter Summary

More children with SVHD are surviving to adolescence and adulthood, but are faced with an uncertain future, with physical, psychological and functional challenges related to their heart disease and its treatment. Adolescents with SVHD are frequent users of healthcare and support services. Thus, healthcare providers must understand and recognize the challenges these individuals face to provide comprehensive and integrative care to achieve their best possible health. In addition, other professionals such as educators and providers of support services should be aware of the challenges these individuals face to provide appropriate support services that facilitate the achievement of their highest functional potential. This study proposes to examine and describe the lived experiences of school, relationships with others, and healthcare encounters in adolescent Fontan survivors. This first chapter serves as an introduction to the problem, purpose, and aims of the study. The second chapter provides a comprehensive review of literature related to the research topic and questions while the third chapter describes the theoretical and conceptual underpinnings of the study. The fourth chapter describes the research methodology and the fifth chapter presents the study results. The final chapter will be a discussion and synthesis of the study findings in the context of the conceptual framework with implications for clinical practice and recommendations for the future.

Chapter 2 Review of Literature

This chapter reviews the relevant literature to frame the research questions presented in this study. The initial sections address the physical and neurodevelopmental challenges that adolescents with SVHD face. These co-morbidities, along with the cognitive, behavioral, executive function, and psychological challenges that are associated with SVHD form the context in which adolescents with SVHD experience school and social relationships as well as healthcare encounters. Next, the literature review focuses on the research questions being posed in this study. Finally, the literature review highlights current knowledge gaps regarding adolescents with SVHD.

Single Ventricle Heart Disease

Single ventricle heart disease is a complex and rare subset of CHD, encompassing a variety of structural cardiac malformations that cannot support bi-ventricular circulation (O'Leary, 2002). There is usually hypoplasia or atresia of one of the atrioventricular valves and/or ventricles, and often there is an obstruction to either the pulmonary or systemic outflow tract. The incidence of CHD is estimated at approximately 1.0% of live births, and SVHD accounts for 7.7% of all CHD (O'Leary, 2002). Extracardiac congenital anomalies and genetic abnormalities or syndromes are associated with 11% to 29% of children with SVHD (Egbe, Lee, Ho, Uppu, & Srivastava, 2014; Patel et al., 2016).

The underlying physiology of SVHD is similar regardless of the anatomic specifics. Mixing of oxygenated and deoxygenated blood results in cyanosis. Heart failure (HF) often develops as neonatal pulmonary vascular resistance falls. If systemic or pulmonary circulation is obstructed, pulmonary or systemic blood flow may be compromised following closure of the ductus arteriosus, resulting in potentially life-threatening multiple-organ injury. Surgical

palliation for SVHD is staged. Neonatal surgical intervention is commonly required to regulate pulmonary blood flow through a pulmonary artery band or aorto-pulmonary shunt, with or without aortic arch reconstruction or Norwood procedure to establish stable systemic blood flow (Peterson & Pike, 2018). In-hospital mortality following neonatal cardiac surgery for SVHD currently ranges from 7% to 16% but varies based on co-morbidities and underlying cardiac anatomy (Heidari-Bateni, Norouzi, Hall, Brar, & Eghtesady, 2014; Petrucci et al., 2011; Tabbutt et al., 2012). The second palliative operation, the bi-directional Glenn anastomosis, is done at approximately four to six months of age to further regulate pulmonary blood flow via a superior cavo-pulmonary anastomosis. In-hospital mortality for the bi-directional Glenn was reported as 9% (Deebis, Gabal, Abdelsadek, & Fadaly, 2017). The final palliation for SVHD is the total cavo-pulmonary connection, or Fontan procedure, performed at approximately two to three years of age. Following the Fontan procedure, the pulmonary blood flow from the vena cavae drains passively to the lungs without a dedicated subpulmonary ventricle, and the systemic blood flow is pumped to the body by the single ventricle (Peterson & Pike, 2018). In-hospital mortality for the Fontan procedure was reported as 15-20% for the early surgical eras but has decreased over time to as low as 5% for patients with favorable pre-Fontan hemodynamics (Driscoll, 2007).

Long-term Survival

The worldwide population of patients with a Fontan circulation has grown to an estimated 50,000 to 70,000 patients in 2018; approximately 40% of these Fontan patients are older than 18 years of age (Schilling et al., 2016). Current estimates of 30-year survival after surgical Fontan completion are approximately 85% (d'Udekem et al., 2014; Khairy et al., 2008; Pundi et al., 2015).

Physical Health Challenges

Fontan survivors face life-long health challenges related to their unique physiology of passive pulmonary blood flow and the sequelae of multiple surgical procedures. Their medical care requires high resource utilization. A study conducted in Canada reported that 87% of patients with CHD received subspecialist care, 68% visited emergency departments, and 51% were hospitalized; those with severe CHD including SVHD had higher utilization rates (Mackie, Pilote, Ionescu-Ittu, Rahme, & Marelli, 2007). Atrial arrhythmias affected up to 75% of Fontan survivors (Pike et al., 2011). Pundi and colleagues (2015) reported that freedom from arrhythmias following the Fontan procedure was 24% at 30 years, and overall, 23% of Fontan survivors required permanent pacemaker placement.

Fontan survivors face chronic hepatic injury due to increased central venous pressure, non-pulsatile venous drainage, and decreased cardiac output, which over time produces some degree of hepatic cirrhosis in almost all patients, now recognized as Fontan-Associated Liver Disease (FALD) (Daniels et al., 2017). Thromboembolic complications (e.g., pulmonary embolus, stroke) were reported in up to one-third of Fontan survivors and attributed to venous stasis, poor ventricular function, and residual right-to-left shunts (Viswanathan, 2016). Heart failure affected 27% of adult Fontan patients and was associated with mortality, need for a heart transplant, or the initiation of palliative care (Egbe et al., 2017). Guidelines for the management of HF in patients with CHD, including SVHD, have been developed, although there remains considerable variability in provider practices (Gnanappa et al., 2017; Stout et al., 2016, 2018).

One particularly challenging but rare sequelae of Fontan physiology is protein-losing enteropathy (PLE), or chronic abnormal protein loss through the gastrointestinal tract (Johnson, Driscoll, & O'Leary, 2012; Rychik, 2007). The lymphatic circulation is also profoundly

adversely affected by chronic venous congestion and is now recognized as an important contributor to PLE, FALD, plastic bronchitis, and other Fontan-associated end-organ diseases (Kreutzer & Kreutzer, 2017). Effective treatment strategies for Fontan-associated sequelae are the subject of current and ongoing research.

Neurodevelopmental Challenges

Neurodevelopmental delays of varying degrees, but generally following similar patterns, are identified in more than 50% of children with complex CHD, including SVHD (Marino et al., 2012). This "neurodevelopmental phenotype" includes mild cognitive, gross and fine motor delays, impairments in social cognition and communication skills, and deficiencies in visual/spatial and visual/motor organization and executive functioning. In addition, this spectrum of neurodevelopmental difficulties includes behavioral issues such as inattention and impulsivity as well as mental health disorders (Kharitonova & Marino, 2016). Risk factors for neurodevelopmental delays include prenatal, preoperative, perioperative, and postoperative factors, some of which are not modifiable beyond a certain extent (Mussatto et al., 2014). Prenatal factors include genetic and neurological co-morbidities, including structural brain abnormalities, alterations in fetal cerebral blood flow, and delayed neonatal neurological maturation (Donofrio & Massaro, 2010; Khalil et al., 2014; Licht et al., 2009). Preoperative factors include cyanosis, delayed recognition of critical CHD leading to multiple-organ injury, and perioperative factors include the use of deep hypothermic circulatory arrest, low cardiac output, seizures, need for mechanical circulatory support or cardiopulmonary resuscitation, exposure to volatile anesthetics, and intensive care/hospital length of stay (Limperopoulos et al., 2002; Marino et al., 2012; Wernovsky, 2006). Postoperative factors include family functioning, maternal education, and socioeconomic status (Majnemer et al., 2006; Newburger et al., 2012).

A growing focus of congenital cardiac care is supporting optimal neurodevelopmental achievement. The importance of providing developmentally supportive care to infants and children with CHD today is driven in part by the neurodevelopmental outcomes of survivors from previous eras who are now adolescents and adults. Guidelines for neurodevelopmental screening, assessment, and referral for all children with CHD from infancy to adulthood have been published (Marino et al., 2012). The neurodevelopmental effects of CHD, especially SVHD, evolve and develop over time (Marino et al., 2012). There is growing multidisciplinary interest in providing family-centered, developmentally supportive care to infants and children with CHD and their families to improve outcomes into adulthood, although research in the effectiveness of nursing interventions on developmental outcomes is in its infancy (Lisanti, Cribben, Connock, Lessen, & Medoff-Cooper, 2016; Lisanti, Golfenshtein, & Medoff-Cooper, 2017; Lisanti et al., 2019; Peterson, 2018; Peterson & Evangelista, 2017; Sood et al., 2016).

Effects of SVHD on Academics and Social Relationships

Literature Search Strategies

A literature review was conducted using PubMed, CINAHL, and PsychInfo. Two primary search terms (Fontan procedure, single ventricle) were individually coupled with a larger number of secondary search terms (executive functioning, cognition, language disorders, learning disorders, behavior disorders, attention-deficit hyperactivity disorder, psychomotor performance, social outcome, psychological outcome, psychiatric illness, anxiety, depression, self-esteem, self-concept, body image, school, academics, school performance, bullying, social relationships, autism, qualitative study, healthcare experiences, patient-clinician relationship, perceived health, functional status, and quality of life). No results were found for “healthcare experiences”. Expanding the “healthcare experiences” search terms to include all CHD also

yielded no results. Expanding the "healthcare experiences" search to all adolescents yielded 3 studies on the use of digital communications in adolescents with chronic illness, but this is outside the scope of this review and laden with ethical and privacy concerns. Filters included publication date range from 2000 to 2019, adolescent age group (13 to 18 years of age), English language, research studies, and humans. The abstracts were reviewed for acceptability to the search criteria, and the references of relevant manuscripts were also reviewed to identify other publications meeting the inclusion criteria. Studies that included only adults older than 18 years of age or only children less than 12 years of age were excluded, as were studies that included only simple or moderate complexity CHD. Because of the lack of published qualitative studies meeting these criteria and because the focus of this study was qualitative, the inclusion criteria for qualitative research were expanded slightly to incorporate manuscripts that included young adult participants. After reviewing abstracts and removing duplicates and manuscripts that did not meet the inclusion criteria, 40 manuscripts (10 qualitative, 30 quantitative) were included in this literature review.

Cognitive Challenges

Cognition, or the process of acquiring knowledge and understanding, is a complex set of processes that are often impaired as a part of the neurodevelopmental delays associated with SVHD. Adolescents with SVHD performed significantly worse than healthy age-, gender, and ethnicity-matched controls on two standard tests of cognition, the Montreal Cognitive Assessment and the Wide Range Assessment of Memory and Learning, 2nd edition (Pike et al., 2018). In addition, structural brain abnormalities in regions associated with cognition were identified in adolescents with SVHD (Pike et al., 2018). A study using brain imaging showed that adolescents who had a Fontan procedure had greater abnormalities of white matter

microstructure compared to healthy controls; these abnormalities were correlated with lower Full-Scale Intelligence Quotient and reduced processing speed (Watson et al., 2018) A large study of adolescents with SVHD reported that the SVHD group scored lower than population norms as well as healthy controls in multiple cognitive assessments including Full-Scale Intelligence Quotient, memory, attention, visuospatial skills, and academic achievement (Bellinger et al., 2015). The brain magnetic resonance imaging from 66% of patients with SVHD had some type of structural abnormality compared to 6% in healthy controls (Bellinger et al., 2015). Another study of 133 Fontan survivors reported decreased overall intelligence and academic achievement compared to population norms, but this study had a very large age range of participants (3.7 to 41 years), so the older patients may have had very different medical care than younger patients (Wernovsky et al., 2000). These studies highlight common cognitive deficits in adolescents with SVHD which can impact academic performance.

Executive Functioning

Executive functioning (EF), or the set of complex skills that allow regulation of behavior, emotion, cognition, and social adaptation, are mediated by multiple brain sites in the frontal/prefrontal, parietal, cerebellar, and subcortical areas; deficits in EF have been reported in adolescents with complex CHD including SVHD in many studies (Cassidy, White, DeMaso, Newburger, & Bellinger, 2015; Marino et al., 2011; Sanz et al., 2017, 2018). The most commonly used measure of EF, the Brief Rating Inventory of Executive Functioning includes subscales of metacognition or working memory and behavioral regulation (Gerstle, Beebe, Drotar, Cassedy, & Marino, 2016). Executive function deficits were associated with decreased psychological QOL as well as lower educational and occupational attainment, although this study included a spectrum of CHD and only 43% had SVHD (Sanz et al., 2018). A study

reporting executive function profiles for children and adolescents with cyanotic CHD, including 30% with SVHD, reported executive function deficits in 75-81% and 43% of participants with CHD vs. healthy controls, respectively (Cassidy et al., 2015). Early term (37 to 38 weeks gestation) birth was also associated with worse executive function than birth at ≥ 39 weeks gestation in adolescents with SVHD (Calderon et al., 2016). In a study of 156 adolescents who had undergone a Fontan procedure, Bellinger and colleagues (2015) reported that patients with SVHD had significantly lower EF scores than normal population norms.

Two of these studies were specific to adolescents with SVHD (Bellinger et al., 2015; Calderon et al., 2016) but the majority of studies included patients with a spectrum of CHD and included some patients with SVHD (Cassidy et al., 2015; Gerstle et al., 2016; Marino et al., 2011; Sanz et al., 2017, 2018). This may limit the generalizability of the findings to the SVHD population, but it is evident from these studies that EF is commonly impaired in adolescents with complex CHD and SVHD. Thus, the lack of EF skills can impact academic achievement and daily life activities including the ability to establish and maintain social relationships.

Behavioral Challenges

The second component of EF is behavioral regulation, and behavioral problems are not uncommonly identified in adolescents with SVHD. A study including 104 patients ages 2 to 20 years with SVHD reported that compared to population norms, the SVHD group had higher levels of internalizing and externalizing behaviors and overall behavioral problems as well as lower intelligence measures (Vahsen, Bröder, Hraska, & Schneider, 2018). Calderon and colleagues (2016) also reported that adolescents with SVHD who were born at 37 to 38 weeks gestation were more likely than those born at ≥ 39 weeks gestation to have attention-deficit hyperactivity disorder (55% vs. 26%). Another study of adolescents with SVHD reported a

higher incidence of attention deficit hyperactivity disorder as well as reported disruptive behaviors compared to healthy controls (DeMaso et al., 2017). Older children and adolescents with a specific type of SVHD (i.e. hypoplastic left heart syndrome) were shown to have a higher incidence of autism or attention-deficit hyperactivity disorder than those with other types of SVHD diagnoses (Davidson, Gringras, Fairhurst, & Simpson, 2015). A study that included younger children (ages 7 to 15 years) and a spectrum of CHD (27% with SVHD) reported higher levels of parent and teacher-reported inattention as well as hyperactivity and impulsivity compared to controls without attention-deficit hyperactivity disorder (Hansen et al., 2012)

The studies described above consistently reported a higher incidence of problem behaviors in adolescents with SVHD. Some studies included a wider age range than adolescence specifically. Potential problems with including other ages include the evolving nature of developmental delays that may not be evident in younger children, as well as differences in medical management strategies that may influence behavioral outcomes in both younger and older patients. Behavior regulation or problems can potentially affect the classroom environment, academic performance, peer relationships and social interaction in adolescents with SVHD.

Psychological Challenges

Psychological challenges are common in adolescents with SVHD. Pike and colleagues (2012) reported that patients with SVHD who were less than 21 years of age had a poorer body image than healthy controls, but there was no difference in self-esteem. A qualitative study of adolescents with a variety of CHD diagnoses reported that females reported not wanting their surgical scars to be visible, because it was an indication of being different from peers (Shearer et al., 2013). Anxiety and depression were also common in adolescents with SVHD. In conjunction

with brain imaging abnormalities, adolescents with SVHD were reported to have higher levels of anxiety and depression than healthy controls (Pike et al., 2018). Adolescents with SVHD also exhibited higher lifetime rates of anxiety as well as higher anxiety and depression scores than healthy controls (DeMaso et al., 2017). Some qualitative studies also describe anxiety and depression. A qualitative study of adolescents and young adults with CHD reported that participants described suffering due to anxiety and uncertainty about being able to remain employed and the long-term outlook for their heart disease, as well as body image concerns related to surgical scars (Chiang et al., 2015). Uncertainty regarding long-term health and potential negative impacts on employment and being able to provide for family caused anxiety and sadness for many participants (Berghammer et al., 2015; Chiang et al., 2015; du Plessis et al., 2018; Lee & Kim, 2012; McMurray et al., 2001; Overgaard et al., 2013). From these studies, it is apparent that anxiety and depression, as well as uncertainty, are common mood abnormalities in adolescents with SVHD. These psychological deficits can affect both school performance and cause withdrawal from social situations or the development of relationships.

Academic Impacts

The complex challenges with physical, neurodevelopmental, and psychosocial health associated with SVHD impact the functional status of the adolescent with SVHD. School is the "business" of children and adolescents and is an important predecessor to later independent living and successful employment. Adolescents with complex CHD, including 41% with SVHD, scored lower in reading and mathematics skills than healthy controls and lower scores in visuospatial skills (Bean Jaworski et al., 2018). A study by Gerstle and colleagues (2016) reported that older children and adolescents with SVHD had lower standardized school grades and required more educational support than children and adolescents with other types of CHD.

A study of Fontan survivors 6 to 18 years of age identified parent-reported difficulty in attention in 46% of children and learning difficulties in 46% of the Fontan survivors (McCrindle et al., 2006). Although the type of heart disease was not specified, children and adolescents with special healthcare needs, 6 to 17 years of age with heart disease, were more likely to have problems with learning and concentration, more likely to miss at least 11 days of school, and more likely to receive special education services than children with special healthcare needs who did not have heart disease (Farr, Downing, Riehle-Colarusso, & Abarbanell, 2018). Mahle and colleagues (2000) reported that in school-age children and adolescents with hypoplastic left heart syndrome, a type of SVHD, 83% of parents described their child's academic achievement as average or above average, but 32% received some type of special education services and 19% were held back a grade level in school. Another study of adolescents with SVHD reported lower reading and mathematics composite scores compared to healthy controls (Bellinger et al., 2015). Finally, a population-based study reported that children ages 8 to 18 years with any type of CHD were more likely to receive special education services than children without CHD, although the number of children with SVHD was very small and medical information on treatment was not available (Riehle-Colarusso et al., 2015).

Another potential factor in academic challenges associated with SVHD is bullying, although the effects of bullying may be more related to QOL and mental health than academic achievement. Adolescents with chronic health conditions have reported bullying more frequently than adolescents without chronic health conditions (Pittet et al., 2010). However, there are no quantitative studies specifically related to bullying in adolescents with SVHD or other specific chronic health conditions. A qualitative study reported that adolescents with a spectrum of CHD (19% with SVHD) reported that 32% of the participants reported being bullied at school, usually

in response to being perceived as different from peers (McMurray et al., 2001). In another qualitative study of adolescents and young adults with a spectrum of CHD (23% severe), some of the participants reported being labeled as different and therefore being a target for bullying (Chiang et al., 2015).

These studies report significant educational difficulties in adolescents with SVHD, although several of these studies also included younger school-aged children as well as other types of CHD, which limits their generalizability to adolescents with SVHD. Some studies report medical risk factors for needing special education services or poorer academic achievement. However, it is not reported whether these academic challenges were primarily related to the cognitive or behavioral challenges associated with SVHD, or more related to other problems such as absenteeism, bullying, or psychological co-morbidities. The incidence of bullying and its effects on adolescents with SVHD is not known; the two qualitative studies that identified bullying included a broad spectrum of CHD. Thus, future studies should focus on academic challenges in the single ventricle population from a qualitative lens.

Social Relationships

Adolescence, in general, is a difficult time when peer/social relationships change from same-sex to opposite sex and coupled with the desire to be attractive to others. Given the difficulties that adolescents with SVHD have with body image, mood disorders, behavior disorders, executive dysfunction, school bullying, and other academic problems, it would be expected that social relationships with parents, friends, peers, and others may be difficult. However, there are very few studies that report on this important aspect of life with SVHD. Social cognition or the ability to "read" other people's emotions and intentions through non-verbal cues was significantly reduced in adolescents with SVHD compared to healthy controls in

one study, as well as their ability to recognize their own emotions, or alexithymia (Bellinger et al., 2015). Bellinger and colleagues (2015) also identified a higher incidence of autism in adolescents with SVHD compared to controls. A qualitative study of adolescents and young adults with a spectrum of CHD reported that some participants discussed "interpersonal frustrations" in peer relationships, usually related to being different from peers (Chiang et al., 2015). Another qualitative study of young adults with a spectrum of CHD reported that participants felt their CHD had an impact on relationships with others in that they were sometimes judged or rejected for disclosing their CHD, or sometimes were overprotected due to perceived fragility (Cornett & Simms, 2014). Another qualitative study of adolescents with a spectrum of CHD including SVHD identified that participants sometimes felt excluded by others due to intolerance, overprotection, or ignorance of strangers and being stared at (McMurray et al., 2001). In several qualitative studies, participants felt safe and secure in relationships with their closest friends and immediate family, but these circles were often small (Claessens et al., 2005; Cornett & Simms, 2014; Lee & Kim, 2012; Overgaard et al., 2013; Shearer et al., 2013; Zahmacioglu et al., 2011).

Social relationships are likely shaped by other neurodevelopmental and psychological consequences of SVHD but are not well studied in this population of adolescents. Certainly, it is a difficult subject to study quantitatively, so qualitative research may be best suited to explore the experiences of social relationships. None of the qualitative studies identified were performed in the United States, and the study populations included younger and older age groups as well as some studies including a variety of CHD.

Healthcare Encounters, Perceived Health, and Quality of Life

Healthcare Encounters

Despite frequent healthcare utilization in adolescents with SVHD, there were no published quantitative studies on the nature of their healthcare experiences and how the adolescent viewed these experiences. There were, however, five qualitative studies that examined experiences with healthcare. One study of adolescents and young adults with a spectrum of CHD including SVHD identified that one concerning aspect of the transition from pediatric to adult cardiac care was the need to re-establish a trusting relationship with a new healthcare team (Chiang et al., 2015). Shearer and colleagues (2013) found that adolescents with a range of CHD were more comfortable and "experienced" with medical treatments than their peers, reducing their anxiety about healthcare encounters (Shearer et al., 2013). A separate study of adolescent and adult Fontan survivors reported that participants felt they had "mastered", or become comfortable with, medical procedures due to their SVHD (Berghammer et al., 2015). Young adults with complex CHD including SVHD reported that having surgery in adolescence or adulthood led to anxiety and insecurity due to lack of knowledge about what was happening to them (Claessens et al., 2005). Finally, Lee and Kim (2012) reported that participants felt a sense of belonging related to the emotional support offered by healthcare providers.

Although not well studied in adolescents with SVHD, it seems logical that adolescents' current and past experiences with healthcare may affect their appraisal of future healthcare experiences, as well as adherence to treatment and other health outcomes. Participants of these previous qualitative studies generally reported feeling emotionally supported and had trusting relationships with their healthcare team, although some felt significant anxiety about surgical procedures later in life.

Defining Perceived Health and QOL

The terms "quality of life," "health-related quality of life," and perceived health are sometimes used interchangeably, but these terms have different meanings that are conceptually important. A definition of QOL from the World Health Organization is "…the individual's perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards and concerns" (WHOQOL Group, 1998). Health-related QOL is generally thought of as a subset of QOL, pertaining to physical and emotional health status. An important but often overlooked aspect of QOL and HRQOL is that their subjective natures mean that there may be developmental implications in defining these terms. A concept analysis of HRQOL in adolescents with chronic illness offered the following definition:

> "HRQOL (health-related quality of life) in young people with chronic illness is subjective, multidimensional and dynamic. It is unique to each individual young person and includes aspects of physical, psychological, and social function. It is dependent upon not only the stage of development but also the illness trajectory. This involves the achievement of goals and aspirations and the constraints imposed through ill-health and treatment" (Taylor, Gibson, & Franck, 2008, p. 1831).

The constructs of QOL and HRQOL are related but not interchangeable, since health may influence overall QOL, but poor health does not always equate to low QOL. Perceived health is a subset of HRQOL that focuses on self-reported functional status or the impact of disease and its treatment on daily life and the ability to function in the context of disease, and the term is sometimes used interchangeably with HRQOL (Moons, 2004).

Perceived Health and QOL

Mean HRQOL scores in adolescent and young adult Fontan survivors were 70-80 (higher scores indicate better HRQOL; maximum 100) (Pike et al., 2011). Compared to healthy controls, adolescent and young adult Fontan survivors reported lower levels of physical perceived health and higher depression scores than healthy controls, but no differences were found between groups in overall QOL, mental health status, and social support (Pike et al., 2012). In contrast, a German study on functional outcomes reported that HRQOL was not different between Fontan patients (mean age 12 years) and healthy peers (Hock et al., 2018). Vahsen and colleagues (2018), also reported no differences between HRQOL in school-age and adolescent Fontan survivors and healthy controls despite significantly increased behavioral problems and visuospatial processing in the Fontan group. Medical history and worse current medical status were predictors of decreased parent- and patient-reported HRQOL in one study of school-age and adolescent Fontan survivors (Dulfer et al., 2016)

The Pediatric Heart Network (PHN) is a large multi-institutional research group that produced high-quality research on many aspects of SVHD. Findings showed decreased physical and psychosocial HRQOL in adolescent and young adult Fontan survivors compared to healthy controls, with 45% and 30% of Fontan patients reporting impaired physical health and psychosocial health, respectively (Uzark et al., 2016). In one PHN study, adolescent Fontan survivors rated their perceived physical functioning marginally lower than population normative values but higher than normative values in many other aspects (McCrindle et al., 2014). However, a previous PHN study reported that parents of school-age and adolescent Fontan survivors rated their child's physical and psychosocial health worse than normative values (McCrindle et al., 2006). Interestingly, another similar PHN study of school-age and adolescents

Fontan survivors reported that parent-reported physical and psychosocial health was significantly lower than population normative values, but that 80% of the scores were within normal ranges, suggesting that some scores were significantly lower (Anderson et al., 2008). In addition, school–age and adolescent Fontan survivors reported that Fontan survivors who had siblings rated their physical functioning and overall QOL lower than those without siblings, suggesting that having a constant comparison to "normal" changes the perspective of physical limitations (Manlhiot et al., 2009).

A recent study of Fontan survivors aged 8 to 18 years showed that parents' ratings of their child's QOL were lower than the child's rating and QOL was associated with abnormal anthropometrics, poverty, and a diagnosis of hypoplastic left heart syndrome, one type of single ventricle diagnosis (Menon et al., 2018). Another study of QOL compared children and adolescents with SVHD to those with another type of cyanotic CHD, tetralogy of Fallot, and reported that the former had lower total QOL based on both parent and patient reports (Eagleson, Justo, Ware, Johnson, & Boyle, 2013).

Qualitative studies have also examined QOL from a different perspective. Zahmacioglu et al. (2011) reported on psychological conditions of adolescents with SVHD and identified an overall theme of "being strong and resistive" by using effective coping mechanisms and social support. Concerns about QOL among young and middle-aged adults were related to potential future adverse health and being able to access psychological support services (duPleiss et al., 2018). Berghammer and colleagues (2015) identified "being committed to life" among participants with SVHD; their cardiac disease encouraged them to make the most out of life and to create meaning. One study of adolescents with a range of CHD defined QOL from the perspective of the adolescent participants as "It's like, how you enjoy life" (Shearer et al., 2013).

Chiang and colleagues (2015) described how adolescents and young adults developed "co-existence" with their heart disease so that the CHD did not define them, and they could pursue opportunities to demonstrate their abilities and strengths. Overgaard and colleagues (2013) reported that six of eleven adolescents and adults with SVHD stated that having SVHD gave them positive life experiences, such as meeting good friends and that they valued life more than those without heart disease.

Even among adolescents with SVHD, studies report some inconsistent results on HRQOL, perceived health, and QOL measures. Some of these differences may be the result of differences in outcome measures or between subject groups. In addition, parents and healthcare providers also rated QOL differently than the adolescent with SVHD. Other non-medical variables are known to affect HRQOL in adolescents in general, including, gender, socioeconomic status, mental health and parental mental health, self-efficacy, social support, and family dynamics (Otto et al., 2017). The effect of many of these variables on HRQOL in adolescents with SVHD has not been reported but has a significant contribution. Overall, self-reported perceived health and QOL are often relatively high in Fontan survivors, despite the challenges they face with cognitive function, executive function, and mental health. Nonetheless, it is important to continue to study the factors that impact QOL in SVHD survivors to promote and develop strategies to maximize self-reported health and QOL.

Qualitative Studies

Qualitative methodologies are important tools to better understand QOL and the experiences of adolescents with SVHD as they cope with the impact of chronic health conditions on their everyday lives. Table 2.1a and 2.1b summarizes qualitative studies performed since 2001 with adolescents and young adults living with varying types of CHD (Berghammer et al.,

2015; Chiang et al., 2015; Claessens et al., 2005; Cornett & Simms, 2014; du Plessis et al., 2018; Lee & Kim, 2012; McMurray et al., 2001; Overgaard et al., 2013; Shearer et al., 2013; Zahmacioglu et al., 2011). The research questions and qualitative methodologies were varied, as were the cardiac diagnoses of the participants. Four studies included only patients with SVHD (Berghammer et al., 2015; du Plessis et al., 2018; Overgaard et al., 2013; Zahmacioglu et al., 2011), although only one study included the adolescent age group exclusively; the remaining three included adults as well as adolescents. Three studies included participants with a spectrum of CHD, from simple to complex, with the complex CHD (including SVHD) ranging from 19% to 33% of the participants; one study included only complex CHD with 10% having SVHD. In two studies, the distribution of CHD complexity was not reported. The sampling in these studies greatly limits the applicability of findings to the adolescent age group, and to adolescents with SVHD. The experiences of adolescents are unique as they strive to achieve self-identity and become independent, and the experiences of those with SVHD, the most complex type of CHD, are likely very different from those with simple CHD. The participants were enrolled from various countries in Europe, Canada, Asia, and Australia. Participant race and ethnicity of participants were not reported in any of these studies, but the distribution of racial and ethnic groups is likely to be more varied in the United States. Other important differences are the healthcare systems; with single-payer national health services more commonly found outside the United States, as well as differences in primary and secondary education systems.

All the studies used in-person interviews to gather data, except one study where an online survey with open-ended and closed questions was used. Face-to-face interviews have the advantage of being able to tailor questions and probes to the responses of the participant, but the disadvantage of taking more time and resources to complete. Surveys enable larger sample sizes

because of the relative ease of implementation but can be slow to accrue participants. In addition, interesting or incomplete responses cannot be probed further in a survey, and follow-up is impossible if the survey is anonymous (Patton, 2002) Common themes across all the qualitative studies included physical limitations due to CHD and coping with these limitations, the importance of peer and family support, use of positive coping strategies and being different from others. These themes are important to explore further in the context of adolescents with SVHD living in the United States, to identify similarities and differences with previous qualitative studies. It is also important to explore their experiences in school and other social relationships because these are areas that may be impacted by the effects and co-morbidities of SVHD and yet also be important in shaping their transition to adult life and independent living.

Knowledge Gaps

Adolescents with SVHD must deal with ongoing, often serious medical and psychosocial sequelae of their heart disease and may have limited ability to cope with these lifelong challenges. Their ability to function in society, including independent living, educational achievement, and employment may be challenged by the medical and psychosocial comorbidities of SVHD. Adolescents with SVHD are also at high risk for discrimination and bullying. The physical, neurodevelopmental, cognitive, behavioral, and psychological adverse effects of SVHD and its treatment are well-researched. All these potential problems impact academic achievement and the ability to establish social relationships, but these areas are much less well studied in the population of adolescents with SVHD. Despite the evidence that adolescent Fontan survivors typically perceive their health and QOL as relatively good, some studies report conflicting findings, so it is important to explore further. Finally, these adolescents are frequent consumers of healthcare and have often complex healthcare needs, so it is important to better understand

their perceptions of healthcare experiences so that healthcare providers can better assist these at-risk adolescents to live the longest and healthiest life possible.

Chapter Summary

The literature reviewed is significant for the overall lack of qualitative studies focused solely on adolescents with SVHD. This is likely due to the relative rarity of this type of CHD which limits the feasibility of single-center investigations in terms of access to the target population. However, the unique ongoing health challenges and high resource utilization of this at-risk group warrants additional research. There are many quantitative studies of physical health outcomes as well as psychological outcomes, HRQOL, and QOL in the adolescent SVHD population. However, there is limited knowledge of how these challenges impact experiences of school and building social relationships, which are crucial for the transition to independence. Knowledge of their healthcare experiences is also important as these experiences may influence their adherence and participation in ongoing healthcare. These questions may be especially suitable for a qualitative methodology. Another knowledge gap identified is the lack of qualitative research in the adolescent SVHD population within the United States. Although qualitative research has been done in other countries, the differences in school and health care systems make it challenging to extend the results of these studies to adolescents with SVHD in the United States.

Table 2.1a. Qualitative Studies on Adolescents Living with CHD Part 1 (Authors Berghammer through Lee)

Study ID	Participants (N, ages, sex, location)	CHD diagnosis	Methodology	Topic and Scope	Themes and Findings
Berghammer et al.2015	N =7, 17-32 yrs, 29% male, Sweden	SVHD following Fontan palliation	Phenomenological hermeneutic	Positive and negative influence of SVHD on daily life	1. Happiness over being me 2. Focusing on possibilities 3. Being committed to life
Chiang, et al. 2014	N = 35, 15-24 yrs, 60% male, Taiwan	All types of CHD, 23% complex.	Descriptive phenomenology	Impact of CHD on daily life, worries related to CHD, kinds of medical resources/support needed, and recommendations for health care.	1. Invisible defects: imperfect understanding 2. Conflict: Interpersonal frustrations 3. Imbalances: loss of self-balance 4. Suffering: increasing anxiety 5. Encounter: meeting needs 6. Coexistence: positive coping strategies
Claessens, et al. 2005	N = 12, 25-40 yrs, 50% male, Belgium	Cyanotic CHD, 33% had SVHD	Grounded theory	What does it mean to live with CHD	Feeling of being different and process of normalization
Cornett & Simms 2013	N = 7, 21-36 yrs, 28% male, United Kingdom	Unspecified but diverse	Interpretive phenomenological analysis	Psychological impact of living with CHD in adulthood	1. A constant, limiting presence 2. The psychological experience 3. Impact on view of the self 4. Impact on relationships 5. Coping strategies 6. Help and support
du Pleiss, et al. 2018	N = 57, 18-51 yrs, 30% male, Australia and New Zealand.	SVHD following Fontan palliation	Thematic analysis	Greatest concerns in adults with Fontan physiology	1. Physical concerns 2. Pregnancy and children 3. Financial concerns 4. Quality of life
Lee & Kim 2012	N = 10, 14-22 yrs, 50% male, Korea	Complex CHD, 10% with SVHD	Exploratory qualitative descriptive	Understand life experiences of adolescents with complex CHD in a sociocultural context	1. Perception of being different from others 2. Feeling of chaos 3. Feeling of a sense of belonging

N = number of participants; CHD = congenital heart disease; yrs = years of age; SVHD = single ventricle heart disease; SV = single ventricle; QOL = quality of life

Table 2.1b Qualitative Studies on Adolescents Living with CHD Part 2 (Authors McMurray through Zahmacioglu)

Study ID	Participants (N, ages, sex, location)	CHD diagnosis	Methodology	Topic and Scope	Themes and Findings
McMurray, et al. 2001	N = 37, 11-18 yrs, 54% male, United Kingdom	Simple to complex CHD, 19% SVHD	Thematic analysis	Understanding of their disease, areas of life affected by CHD, future aspirations, wishes and concerns	1. Coping with the presence of disease 2. Limitations 3. Exclusion by others 4. Discrimination and bullying 5. Life improvement
Overgaard, et al., 2013	N = 11, 16-48 yrs, 45% male, Denmark	Single ventricle physiology, varying surgical hx	Descriptive phenomenology	How young adults with SV physiology experience daily life and cope with physical/emotional difficulties of the disease.	1. Network support 2. Coping with limitations 3. Life conditions and perspectives
Shearer, et al. 2013	N = 10 (22 interviews), 13-17 yrs, 40% male, Canada	Unspecified but diverse	Interpretive description	How adolescents with CHD describe their everyday life and quality of life	1. QOL is how you enjoy life 2. Physical differences 3. Ability to situate CHD in the background or the foreground of situations.
Zahmacioglu, et al. 2011	N = 17 + parents, 12-20 yrs, 53% male, Turkey	SVHD, varying surgical hx	Grounded theory	Psychological and emotional responses to living with SVHD	Overall theme: being resistive and strong as a coping mechanism. Secondary themes: attitudes of family and friends have positive and negative influence on life with SVHD, coping mechanisms are correlated with personality and temperament

N = number of participants; CHD = congenital heart disease; yrs = years of age; SVHD = single ventricle heart disease; SV = single ventricle; QOL = quality of life

Chapter 3. Theoretical Background

The theoretical and philosophical underpinnings for the research are described in this chapter. The research approach and analysis were guided by phenomenology as described by Husserl (Dowling, 2007). The theoretical frameworks of Social Ecology as described by Bronfenbrenner (Lerner, 2002) and Social Ecology of Resilience as described by Ungar (Michael Ungar, 2011) are used to frame the results of data analysis.

Phenomenology

Phenomenology is both a philosophy, originally described by Edmund Husserl (1859-1938), as well as a qualitative research method (Dowling, 2007). The phenomenologic approach seeks to describe lived experiences, as they are experienced by the individual, in order to discover the meaning of these experiences (Englander, 2012). Knowledge is developed when one is able to explore the meanings of the lived experiences and identify the essence or central theme(s) of the experience (Phillips-Pula, Strunk, & Pickler, 2011). There are three types of phenomenological inquiry: 1) transcendental or descriptive phenomenology, which focuses on essential meanings of the lived experience; 2) existential phenomenology, which explores the nature of the reality that binds the phenomenon together; and 3) hermeneutic phenomenology, which focuses on interpretation and interaction between experiences and the reader. The focus of this study is on identifying the perceptions and meanings of the participants' experiences, so the research approach will be one of descriptive phenomenology.

One core characteristic of the phenomenological methodology described by Husserl is a reflection by the researcher to identify and acknowledge *a priori* conscious and unconscious personal beliefs and biases through a process called "epoché", meaning "to suspend judgment" (Dowling, 2007; Husserl, 1970; Moustakas, 1994). These pre-existing beliefs and biases are then

"bracketed" or put aside intentionally to allow the investigator to approach the research with a fresh and unbiased perspective. Other key characteristics of Husserl's phenomenological philosophy include "imaginative variation," or the acceptance of the descriptions of the experiences exactly as related by the participants in order to see the essence of the experience, "intentionality" or the act of being fully present in the situation in order to identify the "noema" (perception) and "noesis" (meaning). The true understanding of the phenomenon is called "phenomenological reduction" (Dowling, 2007; Husserl, 1970; Moustakas, 1994). The result of phenomenological inquiry is the "essence," or that which holds all of the aspects of the phenomenon together (Phillips-Pula et al., 2011). A follower of Husserl, Amedeo Giorgi developed research and data analysis methods specific to descriptive phenomenological methodology in psychology, which are further described in the data analysis section (Giorgi, 2009; Phillips-Pula et al., 2011).

Theoretical Models

Social Ecological Systems Model

A Social Ecological Systems Model was originally developed by Uri Bronfenbrenner in 1979 to describe human development (Lerner, 2002). This model involves four interrelated concepts: 1) *the developmental process* involving a fused and dynamic relation of the person and the context; 2) *the person*, with his/her unique repertoire of biological, cognitive, emotional, and behavioral characteristics 3) The *context* of human development, conceptualized by nested levels; 4) *time*, conceptualized as involving multiple dimensions of temporality including lifespan, family/generational time, and historical (social/cultural influences) time (Lerner, 2002). The inner spheres of influence on an individual's characteristics, such as family, friends, and teachers, represent microsystems. The interactions between microsystems are called mesosystem

interactions; these determine the nature of resources available to individuals. Exosystems are institutional environments where policies and services are designed and developed; these shape the quality of mesosystem and microsystem interactions. The macrosystem is the level in which customs, laws, and cultural practices influence the individual (Ungar, 2011).

A social ecological systems model perspective is appropriate for this study because of the multiple factors that shape the experiences of adolescents with SVHD including biological factors, family influences, developmental concerns and changes, the influence of school and other social institutions, and the sociocultural environment in which these adolescents are raised and are influenced by. The coping mechanisms used by adolescents with SVHD to lead their lives in the context of having a chronic serious medical condition are shaped by all these factors. Social ecological models have been developed to explain many different social constructs.

Social Ecological Model of Resilience

The overall essence or concept identified in this study, "Thriving in the face of uncertainty" is quite similar to a definition of resilience, or the process of adapting well to adversity (American Psychological Association, 2019). For this reason, a resilience model was selected to frame the results and implications of this study. The Social Ecological Model of Resilience was developed by Michael Ungar in the mid-2000s in response to the need to incorporate the influence of environmental or ecological factors to explain resilience as an adaptation of ecological systems rather than an individual's ability to overcome challenges (Ungar, 2018). The social ecological view of resilience is shaped by four principles, including decentrality, complexity, atypicality, and cultural relativity. Decentrality is removing the focus of defining and studying resilience from the individual and their outcome to the availability and accessibility of culturally relevant environmental resources. Rather than expecting adolescents to

innately adapt to their adversity, social ecologies need to supply or advocate for resources that support the adolescent at risk. Complexity is the nature of the dynamic relationships between protective processes and resources and the predicted outcomes, which may also evolve over time. Atypicality refers to the lack of dichotomous right or wrong adaptation responses or the use of resources. For example, an adolescent dropping out of school may appear to be a "wrong" response, but this response may be adaptive in an environment that marginalizes children based on racial-ethnic identity. Cultural relativity is considering all types of cultural and temporal influences on all aspects of development, resources, and responses (Ungar, 2011; van Rensburg, Theron, & Rothmann, 2018). The definition of resilience proposed (Ungar, 2008) is:

> "In the context of exposure to significant adversity, resilience is both the capacity of individuals to navigate their way to the psychological, social, cultural, and physical resources that sustain their well-being, and their capacity individually and collectively to negotiate for these resources to be provided and experienced in culturally meaningful ways (p. 225)".

In a mixed methods study of adolescents in 11 different cultures, Ungar and colleagues (2007) identified seven environmental characteristics that work in tension with one another that create processes that are associated with resilience, including relationships, access to resources, identity, power and control, cultural adherence, social justice, and cohesion. Navigation to these resources was facilitated or impeded by negotiations between individuals or groups such as families or communities and those who are gatekeepers to the resources (Ungar et al., 2007).

An illustration of a social ecological model of resilience is shown in Figure 3.1. Internal factors include an individual's beliefs and values that influence their perceptions of and interactions with their environment. Internal factors also include physical factors such as the

adolescent's current health status and specific physical, emotional, developmental, cognitive, and behavioral co-morbidities of their SVHD. The adolescent functions within microsystems of family, friends, and social networks that also exert norms and values on the adolescent and also supply important coping resources. External influences include exosystems including organizational and community environments such as school, healthcare, and community support systems. At the macrosystem level, legislative, policy, and media are other types of external influences. The adolescent with SVHD is influenced by all of these systems and in turn these systems are also influenced by the adolescent with SVHD.

From a developmental systems perspective, adolescence is a time of physical maturation as well as enormous cognitive and interpersonal development (Balk, 2014). Cognitive growth includes developing the ability to use abstract concepts and think critically, while interpersonal growth includes the development of self-identity and self-concept. This growth occurs in the context of interactions with "ecological niches" or sociocultural environments including family, peers, school, and social media (Balk, 2014). Adolescents with chronic life-threatening illnesses face additional challenges as they must learn to cope with the impact of their illness on all aspects of their lives. Some especially important challenges are the development of self-concept and self-identity while the effects of a chronic illness identifies the adolescent as different from their peers, which also impacts social acceptance and can lead to isolation (Boice, 1998). The development of resilience is the ability to access and mobilize resources from all of the spheres of influence to respond to these physical, interpersonal, cognitive, emotional, behavioral, and spiritual impacts of the chronic illness (Balk, 2014; Ungar, 2012). Resources identified by participants in this study include family support, personal values and traits, being treated "normally" by siblings and parents, social networks of other adolescents with SVHD, teachers

who understood their needs and provided support, Individualized Education Plans (IEP), spiritual activities and beliefs, trusting relationships with healthcare providers, social acceptance by peers, networks of friends, and heart camp experiences.

Chapter summary

The Social Ecological Model of Resilience is an appropriate frame for the results of this study. Adolescents with SVHD have had to cope with the effects of their heart disease on their daily living, and they face ongoing physical, developmental and psychosocial challenges. Their ability to function in society is the result of many intertwining factors of family functioning and socialization, school support, peer support, excellent healthcare, and societal support.

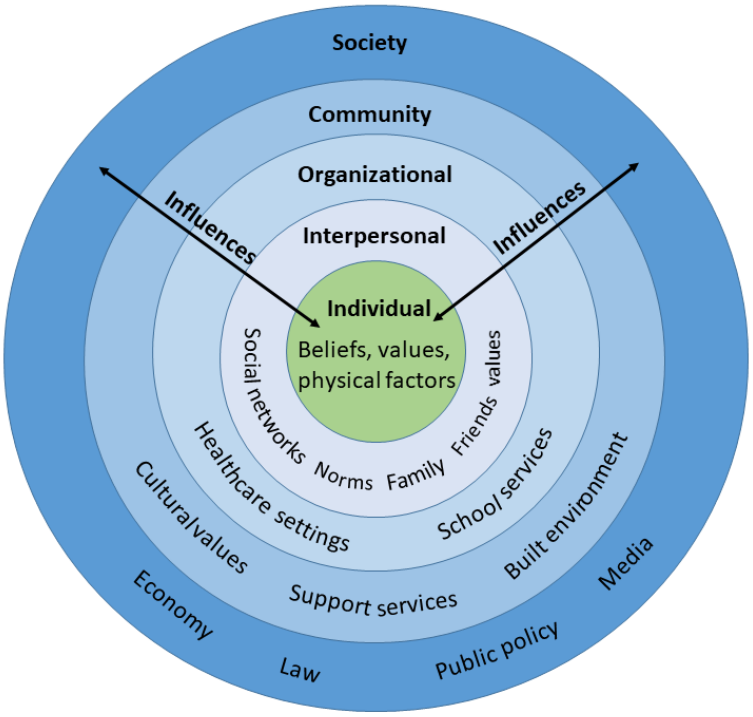

Figure 3.1 Social Ecological Model of Resilience. Adapted from "Psychological resilience" from Local Public Health Institute of Massachusetts.

http://wwwapp1.bumc.bu.edu/lphi/publichealthtraining/onlinecourses/ep-stress/EP-Stress6.html

Chapter 4. Methodology

The purpose of this chapter is to describe the research design and methods used to study the lived experiences of school, social relationships, and healthcare encounters in adolescents who are Fontan survivors. This chapter will review the population, sampling and recruitment procedures, data collection and analysis, as well as issues related to human subjects' protection and the ethical conduct of research. The research questions outlined in Chapter 1 required a methodology that allows for in-depth exploration of perceptions and meanings assigned by participants to their experiences. Among the qualitative methodologies, the descriptive phenomenological approach was selected as most applicable to these requirements because its purpose is to identify the meaning, structure, and essence of the lived experience of a particular phenomenon for a person or group of people (Patton, 2002).

Sampling

Adolescents aged 14 to 19 years of age with SVHD who have undergone a Fontan procedure were purposively sampled. Adolescents were selected because of their current or recent school experiences and developmental capacity for abstract thought, as well as their stage of transition from the care of their parents to independence. The adequacy of the sample size was determined by theoretical saturation of the data, which occurs when the addition of new data no longer results in new categories, insights, or new properties of the core themes (Morse, 1995; Patton, 2002).

Inclusion/Exclusion Criteria

Participants must have had SVHD and previously undergone a Fontan procedure, and be between the ages of 14 and 19 years. The participants had to speak and understand English, as interviews were conducted in English; recruiting non-English participants would require

translation of the transcripts which can potentially impact the context and meaning due to subtle differences in language (van Nes, Abma, Jonsson, & Deeg, 2010). Another important aspect of translation in qualitative interviews is that the translator is not an invisible participant and may also influence the interpretation of the data (Squires, 2009). Participants were excluded if they had received or were currently listed for an organ transplant or if they had a developmental or psychological impairment that prevented them from participating in the interview.

Procedures

Institutional Review Board (IRB) approval was obtained at the University of California Los Angeles before starting the study (#18-001204). Once study approval was received, the principal investigator (PI) met with Dr. Nancy Pike to discuss dissemination to her current and previously enrolled participants from a larger study of adolescents with SVHD assessing cognitive outcomes and brain structure integrity. The following describes the study protocol that was implemented.

Recruitment

The study was conducted through the University of California Los Angeles. The setting was chosen because of the availability of a large cohort of adolescents with SVHD. Participants included those who indicated on their previous informed consent document that they were willing to be contacted regarding participation in follow-up studies, as well as participants who were newly recruited into the main study. Flyers describing the study were sent to potential or past participants of the larger study with contact information to call the PI if interested (Appendix A). Participants were recruited over a five-month period, from September 2018 through February 2019. Previous qualitative studies in this population with similar aims included

7 to 17 participants, so it was generally expected that the sample size would be between 12 and 14 participants (Berghammer et al., 2015; Overgaard et al., 2013; Zahmacioglu et al., 2011).

Screening

Potential participants (or legal guardians if the participant was less than 18 years of age), who responded to the flyers were screened via telephone. The study goals and procedures were explained using a recruitment script (Appendix B), along with the potential risks and benefits. During the phone call, the PI also asked potential participants if they had additional questions regarding the study. For parents of participants < 18 years of age who only spoke Spanish, screening was conducted by a research team member who spoke fluent Spanish using a Spanish recruitment script (Appendix C). If deemed eligible, the informed consent or assent and parental permission documents (Appendices D-G) were sent to the potential participant to review prior to the informed consent discussion. If the potential participant (and legal guardian, if the participant was < 18 years of age) indicated interest in participating, a convenient time and place were arranged to obtain informed consent or assent and parental permission (for participants < 18 years of age) and to complete the study procedures. Although the informed consent document was sent to prospective participants so that they could review it, the actual informed consent and assent procedure took place in person. All members of the research team who obtained informed consent were properly trained to do so. The PI also asked participants to sign a disclosure to use protected health information for research purposes in accordance with Health Insurance Portability and Accountability Act (HIPAA) guidelines (Appendices H-I). The participant, as well as the legal guardian (if the participant was < 18 years of age) were given copies of the signed informed consent document; the original consents were kept by the PI in accordance with the IRB regulations.

Data Collection

Quantitative measures. After obtaining informed consent or assent and parental permission, participants were asked to complete a demographic and clinical information form (Appendix J). Completion of this form took approximately 10 minutes. The demographic information collected was used to estimate socioeconomic status based on four domains: marital status, employment status, educational attainment, and occupational prestige (Cirino et al., 2002; Hollingshead, 1975). This estimation of socioeconomic status classifies parental education into 7 levels from graduate degree to less than the seventh grade and classifies parental occupation into 9 levels ranging from major professionals to unskilled workers. If both parents were employed outside the home, their two scores were combined, and a mean value was calculated. If the parent's primary occupation was homemaking, student, or unemployed or receiving state assistance, then a categorizable occupation was not present (Cirino et al., 2002; Hollingshead, 1975). The final score ranged from 8 to 66, and is a composite of the parental mean score. The scores were then categorized into five socioeconomic strata including lower (8-19), lower middle (20-29), middle (30-39), upper middle (40-54), and upper (55-66).

The demographic form also contained two Likert scale questions on self-perceived health modeled after the Short-Form-36 Health Survey Version 2 (SF-36). The first question queried overall perceived health, with responses ranging from 1 = excellent and 5 = poor, with lower scores indicating better health. The second question queried perceived changes in health over the previous year, with responses ranging from 1 = much better than one year ago to 5 = much worse than one year ago, with lower scores indicating better health. Although single-item scales are sometimes critiqued for lower content validity and sensitivity than multi-item scales, the single-item scale has been found to be acceptable in measuring constructs such as perceived

psychosocial stress, perceived health, QOL and HRQOL (Bowling, 2005; Cunny & Perri, 1991; Haddock et al., 2006; Hoeppner, Kelly, Urbanoski, & Slaymaker, 2011; Littman, White, Satia, Bowen, & Kristal, 2006; Yohannes, Dodd, Morris, & Webb, 2011). Single-item measures can only be psychometrically tested using measures such as test-retest reliability and correlation with other accepted measures. Test-retest reliability for the overall perceived health question in other studies ranged from an intraclass correlation of 0.69 (DeSalvo et al., 2006) to 0.90 (Hart, 2003). In one study, higher self-rated overall health was highly correlated with lower participation in negative health behaviors such as cigarette smoking and binge drinking (Haddock et al., 2006). Negative changes in self-reported perceived health were significantly associated with negative changes in SF-36 scores (Garratt, Ruta, Abdalla, & Russell, 1994).

In addition, a linear analog self-assessment (LAS) was used to measure QOL. The LAS is a vertically oriented line 10 centimeters in length, and participants were asked to indicate which number from 0 (lowest QOL) to 100 (best possible QOL) on the line represented their QOL. The LAS is widely used to measure perceived QOL of life in many populations, including SVHD (Apers et al., 2016; de Boer et al., 2004; Moons, 2005; Moons et al., 2006). As with the single-item self-reported overall health and changes in perceived health, the LAS also cannot be assessed for validity using Chronbach's alpha. However, the QOL LAS has been shown to be well-correlated with other validated measures of QOL, including symptom distress, mood, and mental status in adult patients with cancer (de Boer et al., 2004; Locke et al., 2007; Singh, Satele, Pattabasavaiah, Buckner, & Sloan, 2014).

A medical chart review form was developed by the PI to include specific cardiac diagnosis, current medications, previous surgical and catheterization procedures and cardiologist's interpretation of cardiac function by an echocardiogram (appendix K). The chart

review form was also used to record information about activity restrictions and the most recent height, weight, and oxygen saturation. New York Heart Association functional class was determined from participants descriptions of their activity level and symptoms associated with activity, as well as the cardiologist's classification, if available.

Qualitative interview. The face-to-face interviews with participants were conducted by the PI after completing the initial forms. The interview was held in a private location, using an interview guide developed by the investigators (Appendix L). The interview guide consisted of open-ended questions about how SVHD affected daily activities and relationships, as well as how they perceived their experiences of healthcare and healthcare providers. The questions focused on participants' perceptions of their experiences, their feelings associated with these experiences and what meanings they ascribed to these experiences. Further probing questions were developed to enrich the scope of participants' responses. The interview lasted approximately 25 to 80 minutes, depending on the participant's responses, and was digitally recorded and then transcribed verbatim. The interview guide was pilot-tested during the first four interviews and refined, and then re-submitted to the IRB for approval. Participants could request to review, edit or erase the audiotape of their interview. Erasing the audiotape or transcribed interview by the participant would constitute withdrawal from the study. None of the participants opted to review the transcribed interviews. Two participants participated in a validation interview by telephone to authenticate themes that were identified from the interviews.

Trustworthiness

The scientific merit of qualitative research cannot be evaluated by the usual standards of reliability and validity used in quantitative research because of its subjectivity. Trustworthiness or rigor of qualitative research can be established by ensuring credibility, dependability,

transferability, and conformability (Guba & Lincoln, 1982). In addition, reflexivity is another important component of qualitative research trustworthiness (Patton, 2002).

Credibility

Credibility is a quality similar to internal validity in a quantitative study and represents the degree to which the qualitative data and its interpretation accurately represent the phenomena being studied (Guba & Lincoln, 1982). Credibility in this study is addressed by having researchers with varied clinical and research backgrounds participate in data analysis (investigator triangulation), allowing interview participants to review and edit their interview transcripts if desired, and by going back to interview participants to verify interpretations (member checks). Other strategies to improve credibility in this study included the use of a well-established research methodology, interview tactics to promote honesty, frequent debriefing between the PI and supervisors, and examination of previous research findings to assess congruence (Shenton, 2004).

Transferability

Transferability is the quality of being generalizable to the population at large (Guba & Lincoln, 1982). Transferability may be limited in this study because of the narrow inclusion criteria, but the results should be generalizable to the population of adolescent Fontan survivors who live in an urban setting. Transferability is enhanced by the use of "thick descriptions" which not only describe the behavior and the experience but also their context, making the description more meaningful (Patton, 2002).

Dependability

In a qualitative study, dependability is similar to the concept of reliability in quantitative research, or the stability of the study findings over time. Data interpretation in qualitative data

analysis prevents exact replication, but the study parameters should be well thought out, documented and described so that they are dependable (Guba & Lincoln, 1982). Another method to improve dependability is a detailed description of research design and implementation (Shenton, 2004).

Confirmability

Confirmability describes the degree to which the findings of the study could be confirmed by other researchers, given the same data. Confirmability was enhanced by using an "audit trail" or field notes of the researcher's thoughts about how to proceed with the study, observations, ethical concerns ,and steps taken to manage, analyze and report of data to document the entire research process and how specific pieces of data resulted in specific interpretations (Lincoln & Guba, 1985; Patton, 2002). Audit trails can be used to track and monitor potential research bias during data analysis. In this study, the PI created a chronological index which listed the choices and decisions made, thoughts about participants' behavioral cues and the rationale underlying pattern and category development and identification. The hope is for the audit trail to serve as a basis for other researchers to trace through the steps in data gathering and how conclusions were reached so they can determine whether the study findings can be used to replicate future research. Another important step in enhancing confirmability is the acknowledgment and documentation of underlying assumptions and biases along with rationale for formulating the study in a particular way (Guba & Lincoln, 1982; Korstjens & Moser, 2018).

Reflexivity

Reflexivity is an awareness of the researcher's values, background and past experiences with the phenomenon being studied, and how those qualities influence or bias data collection and analysis (Patton, 2002). This is an important consideration in this study, as the researchers have

extensive clinical experience caring for children with heart disease and their families. Another potential area to consider for reflexively was the relationship between the PI and the participants, and how the participants and PI perceived each other. In this study, the PI did not have any previous relationship with the participants, which required some initial relationship building but also reduced the potential for influence on participants' responses. The potential impact of the PI's experience was carefully considered. The possibility for this inherent bias was mitigated using epoché and bracketing, as well as having several members of the study team, with varied clinical and research backgrounds, participate in data analysis and by using qualitative analysis software. The PI approached the interactions with participants by placing them in the context of gaining new knowledge, since school and interpersonal experiences in the adolescents with SVHD were a relatively unexplored area of research and by placing participants' responses in the context of their perceptions and interpretations. In addition, re-contacting participants was done to verify interpretation and meanings ascribed to the data to ensure that the conclusions were valid in the eyes of the participants.

Data Analysis

Quantitative Data Analysis

Demographic and clinical data, including the perceived health and QOL, were coded and entered and then analyzed using the Statistical Package for the Social Sciences (SPSS) version 25.0 (IBM; Somers, NY). Descriptive statistics including frequencies and percentages were generated for categorical variables to describe the study cohort. Due to the small sample size, non-parametric statistics were calculated for continuous variables such as age and QOL scores.

Qualitative Data Analysis

Audio files from the interviews were transcribed using "TranscribeMe!" a web-based, HIPAA compliant transcription service that is compatible with NVivo® 12 (QRS International Pty Ltd, Victoria, Australia) software. Based on the first four interviews, minor changes were made to the semi-structured interview guide and the changes were approved by the Institutional Review Board. Transcripts were reviewed by the investigators using principles of descriptive phenomenological inquiry to identify themes and meanings related to school experiences, social relationships and healthcare experiences (Patton, 2002; Phillips-Pula et al., 2011). The specific technique was developed by Amedeo Giorgi (Giorgi, 2009). The steps of phenomenological data analysis began with assuming the attitude of phenomenological reduction, with a perspective and sensitivity toward the phenomenon being researched. The PI read through the data in its entirety to get a sense of the whole, then identified meaningful units within the data. Meaningful units, expressed in the participant's words, were transformed by the PI into relevant expressions to identify common subthemes and themes of the experience. Finally, the essence of the experience was expressed from the identified themes (Giorgi, 2009).

Coding or categorizing segments of the interview data was a critical first step in the analytical process and helped the investigator to understand what was happening in the data and the meaning it represented. The use of NVivo qualitative software eased coding and enhanced the validity of data analysis by providing an "audit trail" of meaningful units ascribed to various themes. The identification of meaningful units was done by the researchers within the software package. All the interview transcripts were reviewed, and meaningful units identified by the PI (JKP), and then discussed with the qualitative research consultant (EFO) through regular meetings to guide the development of these transformations into subthemes and themes. The

transcripts were reviewed iteratively, with modifications to the interview guide to enhance and build upon emerging themes. Transcripts were also reviewed by and discussed with the rest of the research team (LSE and NAP) to ensure that coding was congruent between investigators, and differences were resolved through discussion.

To increase the trustworthiness of the findings, two interview participants (one male, one female) who provided especially rich descriptions of their experiences were re-contacted and asked to provide feedback on the themes and subthemes. Their feedback was supportive of the subthemes and themes identified from the interviews, and both participants reported that they were surprised that others shared the same feelings and experiences that they had reported. Based on their feedback, one of the subthemes, relating to the effect that teachers have, was modified slightly to better reflect the participants' experiences.

Responsible Conduct of Research

Ethical Conduct of the Study

The PI completed training on the responsible conduct of research through coursework on quantitative and qualitative research methods, as well as coursework specific to the responsible conduct of research that met the National Institute of Health standards on training required for doctoral programs. The PI also participated in the National Institute of Health Clinical Center Summer Course in Clinical and Translational Research, a two-week program for 30 selected Ph.D. students that included content on the protection of human subjects and the function of an Institutional Review Board. The PI completed Collaborative Institutional Training Initiative coursework required for principal investigators for the protection of human subjects in biomedical research, responsible conduct of research and good clinical practices as required by the University of California, Irvine and the University of California, Los Angeles.

All participants were asked to complete an informed consent to participate in the study, and minors < 18 years old were provided assent in addition to parental permission. This study was designated as "not greater than minimal risk" by the University of California, Los Angeles IRB and received expedited review and approval (#18-001204). Participants who wished to withdraw from the study could do so at any time, and study withdrawal did not have an impact on the medical care they received currently or in the future. Participants who withdrew could elect to erase their audiotaped interviews. No subject withdrew from the study.

Privacy and Confidentiality

All data collection tools and audio files were identified by a unique study number. As the primary risk to participation in this study was a breach of confidentiality, measures taken to reduce this risk are described below in the risks and benefits section.

Risks and Benefits

The potential risks of participation and measures taken to reduce risk were disclosed in the informed consent. There were no anticipated direct benefits for participants who took part in the study, but the results may benefit other adolescents with SVHD and contribute to scientific knowledge. There was a very small risk of inadvertent disclosure of protected health information or breach of confidentiality. All participant data were identified only through a unique study identification number that did not contain protected health information. The link from participant identifiers to the unique study identification number was maintained by the PI in a locked file, separate from other study documents. Following the required maintenance of study records (3 years), all study records will be securely shredded. The interview audio recordings were identified only with the date of the interview and the subject number. One type of breach of confidentiality could occur in the unlikely event that the participant disclosed information that, in

the judgment of the investigator, posed a safety risk to the participant or others, such as disclosure of active suicidality, abuse or intention to harm others. In this case, the PI is a mandated reporter as a healthcare professional and would be required to violate confidentiality. Participants were made aware of this requirement in the informed consent, assent, and parental permission documents. There was a very small risk that participants could develop emotional distress due to interview topics. Participants were made aware that they could choose not to answer any questions that they did not want to answer or that made them too uncomfortable. The PI provided emotional support and compassionate listening throughout the interview to reduce the effect of discussing any adverse memories. If the participant demonstrated continued distress, the PI would have offered to pause the interview briefly to regain composure or to stop the interview completely. If the participant's emotional distress continued and the PI believed that it would be unsafe or not in the participant's best interest to continue, the PI could choose to end the interview. No interviews were ended prematurely, either by the participant or the PI. If the participant indicated a need for additional psychological assistance, the PI would identify an appropriate resource, such as the participant's primary care physician, student health service or school counselor. If the participant required emergency psychological assistance, such as the disclosure of suicidal ideation, they would be escorted to the nearest hospital emergency room for further evaluation. None of the participants requested additional psychological assistance or required emergency treatment. Participants were given a copy of their rights as research participants as outlined by the University of California, Los Angeles Human Subjects' Protection Program.

Payment to Participant

All participants received a $50 cash payment to include the costs of travel and parking as well as the time and inconvenience of study participation. This payment was made after completion of the instruments and interview.

Chapter Summary

A phenomenological approach, described originally by Husserl and then developed into a research methodology by Giorgi, was used to guide this study with research questions related to the lived experiences of adolescent Fontan survivors, and how they perceived and ascribed meaning to how their SVHD influenced their experiences of school, work, relationships, and healthcare encounters. Although participation in this study did not directly benefit the research participants, the potential risks of participation were identified and did not impose an undue burden on the research participants. Appropriate measures to ensure confidentiality and to preserve the rights of research participants were in place.

Chapter 5. Results

Demographic and Clinical Characteristics

A total of 14 participants (seven males, seven females) participated in the study. The participants were predominantly white (64%), ranged in age from 14 to 19 years (mean 16.5 ± 1.7, median 17 years), and all were single and lived with one or both parents. The single ventricle diagnoses were varied, with 64.3% having a systemic right ventricle. Most of the participants (11/14, 78.6%) were from upper middle or upper socioeconomic strata families. Almost all the participants reported some type of school accommodations, including an Individualized Education Plan, extra time to complete assignments and time to take tests, or attendance in Special Education classes. All participants were currently taking at least one daily medication (median 2, IQR 2-5), most commonly aspirin (78.6%) or an angiotensin-converting enzyme inhibitor (71.4%). Four participants were currently taking diuretics. Table 5.1 and 5.2 summarizes the demographic and clinical characteristics of the participants, respectively.

The participants rated their current health highly, with 12/14 (71.4%) responding excellent to good. Compared to 1 year prior, 13/14 (92.9%) thought that their health was the same or better, but one participant reported much worse health than 1 year prior. The median self-reported QOL was 87.5 on a 0 to 100 scale, ranging from 50 to 99.

Interviews

The interviews were conducted by the PI in a private office at the university (n = 4) or at a public library convenient for the participant (n = 10). All the participants completed the entire interview; the length of the interviews ranged from 24 to 81 minutes (median 45 minutes, IQR 38-59 minutes). Twenty-three subthemes were identified from the transcribed interviews. After reviewing 11 interviews, no new subthemes were identified, and the data collected was

redundant of data already collected, so data collection was judged to be sufficient. To further confirm this, three additional interviews were conducted following this point; these interviews provided additional support for the identified subthemes. From 23 subthemes, seven primary themes emerged from the interview transcripts and an overall "essence" was identified and called "Thriving in the Face of Uncertainty." Table 5.3 summarizes the seven themes and related subthemes. The themes included 1) I am learning to take care of myself; 2) I have some limitations, but don't assume: dealing with ableism; 3) I see myself as healthy; 4) I have big plans for the future, but there is also health uncertainty; 5) School can be a challenge; 6) My QOL is very good, and; 7) I have lots of healthcare experiences. Tables 5.4a through 5.4g reports specific interview responses that were associated with each subtheme.

I am learning to take care of myself. Participants were in varying stages of learning the skills and knowledge needed to transition to independent living with a serious chronic health condition, such as taking their medications, knowing their medical history, regulating their activity according to their ability, identifying symptoms of health changes and scheduling/attending medical appointments. They were also learning to advocate for their needs, often by observing their parents. They reported that having SVHD made them more aware of heart-healthy behaviors, such as a healthy diet and exercise to the best of their capability. Several participants described struggling with sadness and anxiety and sometimes were afraid to add to their family's concern for them by disclosing their feelings. Family relationships also figured prominently in their quest for independence. Some adolescents described highly protective "helicopter" parents and a struggle to take on independence. Others described trying to set a good example for younger siblings or support from other family members as they were taking on adult roles. None of the participants had completely transitioned yet to adult congenital cardiac care, although four

subjects were 18 or 19 years of age. Two of the older adolescents described a program run by the local heart camp called Progressive Adult Cardiac Experience (PACE), which is open to 18- to 25-year old young adults and provides information and resources specific to transitioning to adulthood and adult congenital cardiac care.

I have some limitations, but don't assume: Dealing with ableism. All the participants reported some degree of physical limitation related to physical activity endurance, although the degree of their limitation varied with only peak physical exertion in some to impairment in basic activities of daily living in others. Despite these limitations, participants disliked being labeled as "sick" or "fragile" and preferred to set their own limitations because they knew their ability level better than anyone else. Being seen or treated as different or needing special accommodation was a source of frustration. Participants described instances where they were not given opportunities to do certain activities or were treated differently because others assumed they were incapable. They also described a sense of pride and achievement in overcoming physical as well as other types of challenges.

I see myself as healthy. Although they acknowledged some physical limitations and unique physical health challenges related to their SVHD, all but one of the participants described seeing themselves as a healthy person overall. When asked to describe why or why not, they usually ascribed this assessment in terms of healthy behaviors and not physical limitations. This also included having healthy relationships and support systems. The participants were also active in many types of extracurricular and school-based activities typical of adolescents such as band, theater, and Scouts. Many also described finding physical activities that were compatible with their endurance limitations such as golf and yoga. When asked about the meaning of their

surgical scars, most of the participants viewed them as a sign of strength or accomplishment in overcoming a difficult challenge.

I have big plans for the future, but there is also health uncertainty. The participants generally had goals for the future that included careers, family and travel, although several participants acknowledged that their health may impact their future plans. Many had plans for careers in health-related professions. Most identified additional schooling as the major challenge to achieving their goals. The subject of their 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." Participants described anxiety in questioning whether fatigue was "real" or whether they could push through it, and if so, what the consequences might be. Several with early liver disease discussed uncertainty related to long-term liver dysfunction known to affect most Fontan survivors. Others talked about perhaps needing a heart transplant, having a shorter lifespan or even death. These long-term health concerns were a source of worry, and many felt unprepared to face these concerns. Despite these valid concerns and the potential somber implications, the participants were quick to identify that their SVHD did not define them, although it was a source of their uniqueness.

School can be a challenge for me. Participants often described challenges associated with physical education class, sometimes related to being identified as different because of prescribed activity limitations, and sometimes related to not having their limitations taken seriously. 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. A concerning finding was that five of 14 (36%) participants reported being bullied at school, sometimes to the point of needing to change schools. 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 by being available to help with homework questions or recognizing that students were not feeling well but could also sometimes be the source of additional problems. Being identified to other students as needing accommodations was embarrassing, and some participants perceived that teachers sometimes refused to accommodate their needs for additional time for assignments or physical activity limits. Another set of challenges related to impacts of SVHD on school success. Psychological co-morbidities of SVHD such as autism, attention deficit hyperactivity disorder, as well as difficulties in focusing and retaining information complicated school success in some of the participants. Almost half of the participants reported having an Individualized Education Plan in place, with accommodations to allow additional time for tests and assignments, attending Special Education classes, and other accommodations (Table 5.1). In addition, some participants reported missing out on learning related to absenteeism due to illness as well as frequent restroom use related to medications. Four participants reported moving to a private school or were homeschooled due, at least in part, to the effects of the SVHD or being unable to succeed in public school.

My quality of life is very good. Despite the many challenges of living with SVHD, participants rated their overall QOL highly, in both narrative comments and the linear analog scale (Table 5.2 and Table 5.4f). Some participants acknowledged that perhaps their physical health was not ideal, but that overall, they were quite satisfied. One participant stated, "Some days are really good, and some days not so good, but overall it evens out to about average." Participants cited faith as well as increased awareness of the value of life as some ways that their QOL was better than people without heart disease. Participants also commonly identified that SVHD had very

little, if any, influence on their QOL. However, some participants who had current medical concerns related to the effects of the SVHD described more negative effects in quality of life, such as sadness or frustration at not being able to do activities that they had previously enjoyed.

A crucial determinant of QOL was the support systems that participants relied on to help cope with SVHD and its impacts on their lives. These included mostly family and friends, as well as spiritual beliefs. Family and peers that provided unconditional acceptance and support, because they had witnessed the participants' challenges and struggles, were a cornerstone of coping. Another very important source of support was summer heart camp, which many of the participants had attended several times. Participants spoke of the support provided by camp staff and feeling "safe" there because of the medical background of the staff. Many participants spoke about lasting, deep friendships formed with other campers, made even more deeply bonded by the shared experience of living with heart disease. The positive influence of heart camp and the resulting friendships was very apparent; one participant even became tearful in describing how important and affirming the camp experiences were. One of the participants who was asked to provide feedback on the themes reported that having good and reliable support systems was the primary reason for being successful in living with SVHD and having a good QOL.

I have had a lot of healthcare experiences. Participants were asked to describe both positive and negative experiences with healthcare, as well as to provide advice for healthcare providers new to working with adolescents with SVHD. Most of the participants had no recollection or only vague memories of being hospitalized following their initial three cardiac surgeries. Some of the experiences they related were recent, others were in the past. Some told stories about previous hospitalizations that were relayed to them by their parents.

The positive experiences described by the participants were related to longstanding trust in their pediatric cardiologist or cardiac surgeon as well as long-term relationships with others involved in their care. In addition, being treated as an equal or being seen as more than a patient with heart disease was related to positive experiences. Participants recognized a wide variety of professions as being associated with positive experiences, including physicians, nurses, child life specialists, and office support staff. Negative experiences were sometimes related to painful or otherwise unpleasant sensations experienced during medical care. However, participants also described negative experiences related to perceived uncaring or dismissive responses or perceived errors by healthcare providers.

When asked about advice that they would give healthcare providers who were caring for other adolescents with SVHD, most of the responses were related to ensuring that the patient understood and was engaged in the conversation, active listening, as well as humanizing the experience. Being honest, forthcoming, and open to questions was viewed as important, but these adolescents also wanted to be recognized as active participants in medical discussions and treatment decisions.

Word Cloud

A word cloud figure was generated from the supporting quotations that were gathered from the themes and subthemes (see Figure 5.1). This figure shows the prominence of hopeful words expressed by participants such as “can,” “quality” and “life”. The largest representations of the figure are the words “heart”, “life”, and “condition”. Other common words included “school”, “medicine,” “tired”, “doctors” and “little.”

Chapter summary

Fourteen participants (aged 14 to 19 years) were included in the study. All were single and living with one or both parents, from varied socioeconomic backgrounds although a high percentage from upper middle to upper socioeconomic strata. Almost all (12/14) reported some type of school accommodations. The specific cardiac diagnosis also varied, but all were taking at least one daily medication. Self-reported health was good, very good, or excellent in 12/14 participants, and the median self-reported QOL was 87.5 on a scale of 0 (worst possible) to 100 (best possible). Analysis of the interview transcripts identified seven primary themes, including 1) I am learning to take care of myself; 2) I have some limitations, but don't assume: dealing with ableism; 3) I see myself as healthy; 4) I have big plans for the future, but there is also uncertainty; 5) School can be a challenge for me; 6) My QOL is very good: and 7) I have had a lot of healthcare experiences. Overall, these themes resulted in an overarching concept of "Thriving in the face of uncertainty".

Table 5.1. Demographic Characteristics of Study Participants (N = 14)

Characteristic	Results
Age (years) median, (IQR)	17 (15, 18)
Male gender N, (%)	7 (50.0)
Race/Ethnicity N, (%)	
White	9 (64.3)
Hispanic	3 (21.4)
Black	2 (14.3)
Current school grade N (%)	
8th or 9th	2 (14.3)
10th or 11th	4 (28.6)
12th	5 (35.7)
High school graduate	3 (21.4)
Single marital status N, (%)	14 (100)
Lives with N, (%)	
Both parents	11 (78.6)
One parent	3 (21.4)
Siblings	9 (64.3)
Extended family	2 (14.3)
Socioeconomic status strata N, (%)	
1 Lower	0
2 Lower middle	2 (14.3)
3 Middle	1 (7.1)
4 Upper middle	7 (50.0)
5 Upper	4 (28.6)
Health Insurance Type N, (%)	
Public (Medicaid, CCS)	6 (42.9)
Private (HMO, PPO)	7 (50.0)
Unsure	1 7.1)
School Accommodations[1] N (%)	12 (85.7)
IEP	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	1 (7.1)
Type of school N (%)	
Public school	10 (71.4)
Home schooled	2 (14.3)
Private school	2 (14.3)

Given are frequency (N) and percent (%) for categorical variables, median and interquartile range (IQR) for continuous variables. CCS = California Children's Services; HMO = Health Maintenance Organization; PPO = Preferred Provider Organization; IEP = Individualized Education Plan; PT = Physical therapy; OT = Occupational therapy. [1]Type of school accommodations add up to more than 100% because most participants reported more than one type of accommodation.

Table 5.2. Clinical Characteristics of Study Participants (N = 14)

Characteristic	Results
Cardiac Diagnosis N, (%)	
Hypoplastic left heart syndrome	4 (28.6)
Unbalanced AVSD, heterotaxy	3 (21.4)
Double outlet right ventricle	3 (21.4)
Pulmonary atresia, intact ventricular septum	1 (7.1)
Double inlet left ventricle	2 (14.3)
Ebstein's malformation	1 (7.1)
Single ventricle type N, (%)	
Right ventricle dominant	9 (64.3)
Left ventricle dominant	5 (35.7)
Number of cardiac surgeries median (IQR)	3 (3,3)
Fontan type N, (%)	
Extracardiac, non-fenestrated	7 (50.0)
Extracardiac, fenestrated	2 (14.3)
Lateral tunnel, fenestrated	5 (35.7)
NYHA functional class N, (%)	
1 No limitations of physical activity	1 (7.1)
2 Slight limitations of physical activity	10 (71.5)
3 Marked limitation of physical activity	3 (21.4)
4 Symptoms at rest	0
Number daily medications median (IQR)	2 (2, 5)
Activity restrictions N (%)	
Self-limit	9 (64.3)
Physician activity restriction	3 (21.4)
No restriction	2 (14.3)
Self-rated current health N, (%)	
Excellent	3 (21.4)
Very good	8 (57.1)
Good	1 (7.1)
Fair	2 (14.3)
Poor	0
Health compared to one year ago N, (%)	
Much better	2 (14.3)
Somewhat better	5 (35.7)
About the same	6 (42.9)
Somewhat worse	0
Much worse	1 (7.1)
Overall quality of life (0-100) median,(IQR, range)	87.5 (76.2-91.2) (50-99)

Given are frequency (N) and percent (%) for categorical variables, median, range and interquartile range (IQR) for continuous variables. AVSD = atrioventricular septal defect; NYHA = New York Heart Association; PDE-5 = phosphodiesterase-5; ACE = angiotensin converting enzyme.

Table 5.3. Thriving in the Face of Uncertainty themes and subthemes

Theme	Subthemes
1. I am learning to take care of myself	Advocating for my needs My health behaviors Emotional health Family relationships Transitioning to adult cardiac care
2. I have some limitations, but don't assume: Dealing with ableism	Physical limitations Being labeled Being different Overcoming challenges
3. I see myself as healthy	I see myself as healthy because… My extracurricular activities Finding physical activity right for me What my scars mean to me
4. I have big plans for the future, but there is also health uncertainty	Goals for the future Challenges to meeting my goals The elephant in the room: my future health Single ventricle heart disease doesn't define me
5. School can be a challenge	Physical education class Being bullied Teachers can be a big help or another challenge Single ventricle heart disease impacts school sometimes
6. My quality of life is very good	Support from family, friends, and heart camp Describing my quality of life
7. I have lots of healthcare experiences	Positive experiences Negative experiences My advice for healthcare providers

Table 5.4a. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
1. I am learning to take care of myself	**Advocating for my needs** • "…there might have been a couple times where my Mom had to talk to my teachers because I was trying to communicate that I was too tired…" • "I really kind of ask for everything on my own" **My health behaviors** • "You should know what you have or all the medications you take" • "I definitely get exercise at least five days a week" • "It makes me more mindful of what I should eat and what I shouldn't eat" • "I used to not really do it [take my medicine] but now I understand that I actually have to do it. And so I've been doing really good actually. ..I refill on Sundays because I have a container that says the days. And then I have two containers because I take a lot of medicines." • "In reality I'm the only one that can know if something is wrong. I feel it." • "I have to remember to take my morning pill. I forgot what it's called, but it's a white, small one and my Dad gets mad at me if I don't take it" **Emotional health** • "I just don't like people to always know when I'm struggling…[I] make sure I can show everyone that I'm not fragile and everything's OK. And I don't want to add another thing for other people to worry about." • "The anxiety and depression, that'll probably come and go as I go throughout life, but it's knowing that I'm eventually going to come out of that." **Family relationships** • "They have that 'helicopter' parenting style, specifically with me….they're always watching to see if I'm tired…I feel like I don't really need that hover as much as I did maybe when I was a kid." • "My Dad has always been better about relenting and being like 'We need to let her live her life'." • "…a lot of my family will look up to me for being so strong through all of it. Because…it's a lot to go through…" • "I really try my best to be someone for them [younger siblings], someone for them to look up to." • "He [brother-in-law] tells me, 'You know what? If you want people to see you as an adult, stop acting like a little kid. Start maturing up'." • "I think they [parents] would be overprotective, even if I didn't have a heart condition." **Transitioning to adult cardiac care** • "I'm going to stay with him [pediatric cardiologist] until he discharges me to an adult cardiologist. I don't know how long that will be. But I know that it's eventually, most likely, going to happen." • "…as soon as I turned 18, it's like life hit me like a truck. Now I have a bank account, ID, everything. And it's like 'Oh, shoot. Now I'm getting mail'." "…the biggest part of PACE is helping kids who are first-generations to be able to make the transition. There's not an abundance of people who have been dealing with adults with congenital heart defects".

Table 5.4b. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
2. I have some limitations, but don't assume: Dealing with ableism	**Physical limitations** • "I have a placard so I can park a little bit closer to the school." • "My mental level and my physical level are very different. My physical level is needing me to stop, but my mental level can go another mile." • "I can go a million miles but only for a very short amount of time." • "I do get tired more easily, but I can do pretty much anything, honestly." • "Ever since I was little, I could… never keep up with the other kids." • "I'm not allowed to play contact sports is the only things I'm not allowed. It's not a problem, I'm not playing sports." • "When I stopped doing PE, the effect of my heart defect really went away because the only real effect it had was that I got tired faster than others." **Being labeled** • "I don't want the label of 'She's the sick one'." • "…sometimes I like to pretend I don't have one [heart defect] for a little bit. Just to see what other people see in me besides that condition." • "…they're treating me like I'm fragile…" • "…'she's the girl who has the heart condition', or 'she's the girl with some type of medical issue'." • "So those labels, to me, were always very frustrating because I was able to see the positivity in the condition, and I was able to see past the condition which not everyone who knew me was able to do." • "…they wouldn't make fun of me, but just like, I just hear 'he has a heart condition. He has a heart condition'. I'm like, 'Yeah, I do'." • "…they're like, 'Oh, don't do anything. You're too delicate…Don't worry. I'll do it because you can't. You have a heart condition'." • "I got a lot of people saying what I can and can't do, but I mean, they don't even fully know me." **Being different** • "I'd rather go through a few days or a few weeks of feeling horrible [after a heart transplant], and then the rest of my life feeling great, compared to every single day feeling 'I'm tired and I can't do anything'." • "I very much felt like I had been a burden to her because of something out of my control." • "…if you just assume, then you're going to make the person feel like you don't care about them." • "…and I typically don't like people to know I'm on accommodations." **Overcoming challenges** • "Always striving to keep up with everyone else is its own challenge in school." • "I prove people wrong with stuff a lot." • "I think my biggest success so far is just not letting myself get pushed around." • "I realized I wasn't really going to play other sports at the school, and I chose golf….But I stayed persistent and kept trying at it because it's something I wanted to do and it was an outlet that I could do….And then my third year, I finally made the team."

Table 5.4c. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
3. I see myself as healthy	**I am healthy because…** • "My heart is basically all types of screwed up, but it works. So at least I have that". • "We try to eat the healthiest we can, especially with my condition, I need it most of all. I can't just go into Jack in the Box or eat anything." • "I haven't been to the hospital for medical reasons since the third grade." • "…between walking everywhere and all the volunteer work I do, I have a very healthy life." **Extracurricular activities** • "I do Scouts….I've done the program since first grade, since I was able to join Cub Scouts." • "I have a hobby. I like doing comedy sketches or film stuff. And I like the idea of making a movie…" • "I'm one of the editors of my [school] newspaper." • "I do tech work for my school theater…You call the lights, you call the props, you make sure that music goes on when it should. It's a lot of work." • "…she [my Mom] put me in the LAPD Cadet [Explorers]. It helped change me a lot. I found a few friends there." • "…I have robotics…Not many people can say that they've built a computer from scratch." • "…the television network that goes around school, and I'm one of the lead anchors…" • "I have band practice…I'm mainly in percussion. But right now they have me playing piano…" **Physical activities** • "I remember my cardiologist saying about how Fontan patients can do about 70% of what a normal person can do." • "I used to be a dancer. For six years I did competitive dance. So I would just take more breaks. But, yeah, it didn't really affect my ability." • "…one of the teachers started a yoga class, and so I took that up." • "And when I realized that I couldn't do as many athletic sports and whatnot, I went out and found a sport I can do." **What my scar means** • "It looks cool….It's like a memory, kind of." • "It makes me feel unique." • "Ever since I was little, I've been pretty proud of it." • "It means I survived something." • "A sign of courage, I must say, for all the tough things I've been through." • "Not much, other than I've had surgery." • "It means a lot to me. The battle scars, that's what they mean to me, that I've conquered more than the average person…"

Table 5.4d. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
4. I have big plans for the future, but there is also health uncertainty	**Goals for the future** • "I'd like to be working…as a child life specialist. Hopefully in some type of relationship… Being in good health and living life wherever it is that God is putting me. I want to make sure that I'm there." • "I'd like to be in the medical field in some way." • "A heart surgeon." • "I want to travel, just alone, for a little bit...Everyone's saying that I can't, but I want to live on my own." • "I want to work in the entertainment industry." • "I want to change the world. Stuff kind of sucks right now, and I really want to change the world." • "My dream's been to open up my own business, like have my own body shop, be a master mechanic." • "As long as I set my mind to it, I can accomplish anything in my life." • "I want to be a kinesiologist or a team doctor for a professional sport, so something like that because I can't play the sport." • "…go to college, and study abroad in Korea…then maybe possibly-soon, I'll find love, and then I can get a job…something with teaching." **Challenges to achieving my goals** • "…probably the getting through college thing." • "Maybe I could see myself having more health issues." • "Definitely school. That's one of the challenges. That's going to be hard." **The elephant in the room: Future health** • "So if I'm like, 'OK, I'm tired,' it's like, 'But am I really tired, or can I keep going'…If it's real that I can't go any further but I keep going further, I recognize that there would be bad outcomes to that." • "…I have a huge fear of the unknown of what will happen…I don't know if I'll have to have a heart transplant at some point or if things will get worse…I don't know what my life expectancy is either..." • "Considering I have my heart condition…That might limit my lifespan." • "I know why. They can't just give me a medicine that will fix everything. That's why I'm waiting to be put on the transplant list." • "So as I've gotten older, it's causing some more issues with my liver because of the pressure put on it." • "Well they have told me that I'll probably have to get a heart transplant, and if I don't, that I'll die sooner. That's kind of scary to think about, so I don't think about it that much…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." **Single ventricle heart disease doesn't define me** • "I wish I was just normal. I wouldn't have to deal with all of this. But then again I wouldn't be me if I didn't go through all this either. So, yeah." • "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." • "I never made a big deal out of it. It was just me."

Table 5.4e. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
5. School can be a challenge for me	**Physical education class** • "I remember kids in PE being like 'Why don't you have to run the mile?' • "In PE, I actually ran a mile. And I got sent home in a wheelchair because I was so exhausted." • "…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." • "My doctors had actually talked to the school, so it's not like he [PE teacher] didn't know... Apparently, he just didn't seem to care." **Being bullied** • "I would hear them whispering, 'Hey guys, when we go, run really fast so XX doesn't get a seat at the library.' Because they knew that I needed to sit down because walking that distance for me, it was a lot…." • "Every time they got a chance, they would always pick on me, whether it was holding me down with a finger and say, 'Try and move'. Literally, hold you down, just say bad things." • "…they would call me the midget with the heart condition…" • "…I was getting bullied from these kids, they will still see me as the weak one…[my Mom] would see me crying at home… We tried moving schools. That helped out a little bit." • "The district didn't want to do anything for me. And there was some bullying. So they said they'll pay for anything [moved to different school]…Everyone finds something to pick on." **Teachers can be a big help or another challenge** • "They [teachers] would like to think that I was using my heart condition as an excuse to not do things" • "…he [my teacher] called me out…he said, 'did you fill out those cards?'…And then he looked at the class and said, 'If you have special needs or a special situation, you fill out a card for time and a half.' And I was really embarrassed…" • "She [a favorite teacher] moves a lot of stuff around for me. She's constantly asking me, 'What can I do to make things easier for you?'" • "…my teachers get upset at me for having to go to the bathroom more." **SVHD can impact school** • "…before we knew that I had autism... Elementary was just bad for me…Getting in trouble a lot, kicking, doing all sorts of stuff I shouldn't be doing. Almost getting suspended multiple times." • "I hate having IEPs." • "…I have to use the restroom more because I take a medication…." • "I would miss a lot of days because I have to go to the emergency room a lot…. And I got really behind, and they didn't give me my homework, and it was a whole thing." • "Sometimes I have a hard time retaining information from my teachers." • "…I had this honors AP class, and I was failing all my classes, because I would only be able to go to school for two hours, and then I'd call my Mom and I'd say, 'Come pick me up' and I'd be pale and exhausted, and I'd look like I just ran a mile."

Table 5.4f. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
6. My quality of life is very good	**Support from family, friends, and heart camp** • "We treat each other like brothers normally do, by beating the living hell out of each other." • "I've always just really believed that I'm going to be OK. God's got me." • "…someone who can just understand what it is and know that I'm not over-exaggerating things. They just kind of get it." • "…I think they [friends from heart camp] really are the best thing because they would understand, because they all have heart conditions." • "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." • "…they [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..." • "…they [heart camp] taught me not only what my heart condition was, but how to live with it." • "They [heart camp] make you feel so normal there. And they make you do-push you to your limits….I would always cry when I had to go home." • "Camp introduced me to people like me, as well as growing my confidence." • "…they [PACE program] talk about doing stuff on your own. Kind of the transition to starting to live on your own…How to take care of your medicine on your own." **My quality of life** • "In general, I think I have a very good quality of life." • "[My quality of life compared to others is] Better, being that I have God. I have a good outlook on my heart condition. Worse [compared to others], in that I get sick." • "I think in a main way, that it's [my quality of life compared to others] better because I have more of an understanding of how valuable life is." • "I wouldn't say that it's [my quality of life] bad. But there are a few things that make it go down from a 10 to a little less…So it's not as bad as people think. So yeah, like eight and a half, nine." • "I'd rate it like a 65 to 70. Not necessarily anything pertaining to my heart condition, but just generally a pretty average life…Compared to other people, it's probably around the same." • "…the quality of life, unfortunately, is not that great as far as health is concerned." • "Probably about the same [my quality of life compared to others]. There's really nothing that brings me down or makes me super sad about having a heart condition. It's usually a benefit… It gets me out of hard stuff like physical education." • "Very high. I'm super privileged and I live a good life…I don't think my quality of life is any worse because of my heart."

Table 5.4g. Thriving in the face of uncertainty: Themes, subthemes and supporting quotations

Theme	Subthemes and supporting quotations
7. I have lots of healthcare experiences	**Positive experiences** • "They've been my doctors for a long time now, especially cardiology doctors. So I feel like I can trust them…I'm not as scared because I have the trust." • "…the hospital I went to was actually like-they made it kind of fun." • "So they [the nurses] just kind of made it feel like they were just chilling at my house. They didn't make it feel like it was a hospital thing." • "…the receptionist-her name is Maria- she's been there for years since I was little, and she's always really nice and we get to talk and that makes it better." • "I was playing with some Play-Doh….my cardiologist was doing rounds…Him and my surgeon…they came and sat and played Play-Doh with me." • "I just feel really safe…because I trust them, and I trust that they can help me…" • "…there was a child life specialist there with me…and when the needle went in, she saw that I was starting to panic…she immediately started distracting me." • "In general, I've had nurses who-they recognize me when I show up at the hospital." **Negative experiences** • "I was mad that they made me do this because it caused me pain because I had just had my chest ripped open. And I remember it hurt" • "Some doctors-they don't want me. I'm too difficult for them with all of my health issues…And she [the physician], was saying 'How come they assign me so many difficult people?' And 'Not you, not you.' It's like, it is me, totally. I'm difficult." • "She just didn't think it was a big deal. So I ended up going to the hospital and being admitted and it was a whole thing." • "I realized that everything that they told me at my old hospital…that it wasn't true and that they were lying to me about basically everything." • "I'm taking up to 18 medications a day. They'd be like 'Nope. Nope. Just suck it up. Do it. Deal with it'." • "My Mom left me to go home and take a shower. And I was young, so I was crying. And the nurses weren't listening…I was just trying to have somebody next to me or something. …they shut my door and hollered 'Shut up'." • "…they were taking a tube or something out of me and the doctor said 'It's not going to hurt.' But, instantly, as they were taking it out, it hurt me." **Advice for healthcare providers** • "Make sure that they listen to everything that the patient asks. To dumb it down, because doctors will often use big words" • "Kids, once they see that you talk about those things [the child's favorite thing], they see you as a human, versus as this scary person that's going to poke needles in them and make them take horrible medicine." • "It was frustrating growing up because the doctor would ask my parents all the questions, but I knew the answers because I knew what was going on" • "One thing I hated was when doctors and nurses would try to talk about things that they were going to do to me without me being in the room, because that made me feel like it's such a horrible thing that they didn't want to talk about it in front of me." • "Make sure they're [the child with SVHD] happy with who they are." • "…if you…hurt us because it's your job, not that you want to, and we say we hate you, we don't….we just don't like you in that moment because you're hurting us…."

Figure 5.1. Word Cloud generated from supporting quotations from subthemes

Chapter 6. Discussion

This study describes the lived experiences of adolescents with SVHD. Participants shared richly detailed experiences related to their perceived health and QOL, the impact of SVHD on school, and social relationships with friends, family, and teachers, and experiences with their healthcare providers. Previous qualitative studies have included participants from Europe, Australia, New Zealand, and Asia. This study adds perspectives of a more culturally diverse sample, as well as the influence of healthcare and educational systems in the United States on the experiences of Fontan survivors.

Perceived health and QOL

Quantitatively, these adolescents reported high levels of perceived health and quality of life, in the face of SVHD and its associated health problems. All the participants had highly complex heart disease, were on daily medications and had previously undergone multiple cardiac surgical procedures. Despite this, 12 of 14 participants were in New York Heart Association functional class 1 or 2, and most were advised only to self-limit their activities. Their self-reported health and QOL is congruent with most studies that have reported high levels of self-reported health and QOL in adolescents or adults with SVHD (Bordin et al., 2015; Hock et al., 2018; Kukreja et al., 2015; Pike et al., 2012, 2011; Pundi et al., 2015). However, Uzark and colleagues (2016) reported that 45% of adolescent and young adult Fontan survivors reported significantly impaired physical health and 30% reported significantly impaired psychosocial health. They also reported a significant association between physical and psychosocial status and QOL; so, a potential reason for the different findings may be that most of the participants in the present study had little functional impairment. The “disability paradox” has also been proposed to explain high self-reported health and QOL in patients with complex CHD or SVHD. This idea

proposes that people with chronic health problems view their health and QOL differently than those without disability by socially constructing the impact of their chronic illness on daily life. The ability to adapt to physical limitations and be able to accomplish what one wants to do leads to a different frame of reference for defining health and QOL (Albrecht & Devlieger, 1999; Nazli, 2012). In addition, most of the participants in this study were from upper middle or upper socioeconomic status strata, which also may have positively impacted their self-reported QOL (Otto et al., 2017).

Qualitative themes

We identified seven major themes from the perspectives and lived experiences of adolescents with SVHD, contributing to the overall "essence" of thriving in the face of uncertainty. These themes were 1) I am learning to take care of myself; 2) I have some limitations, but don't assume: dealing with ableism; 3) I see myself as healthy; 4) I have big plans for the future, but there is also health uncertainty; 5) School can be a challenge; 6) My QOL is very good, and; 7) I have lots of healthcare experiences. Although other qualitative studies asked slightly different questions, there were many similarities in the findings, as well as some differences. An expanded description of each theme is provided in the subsequent sections.

I am learning to take care of myself

The participants in the present study reported being concerned about maintaining healthy eating and exercise habits, which was one of the criteria they reported in describing themselves as healthy. In a large qualitative study of adults with SVHD from Australia and New Zealand, half of the participants reported exercising at least three days per week (du Plessis et al., 2018). However, nearly half of the participants in Chiang and colleagues' (2015) study of adolescents

and young adults with a spectrum of CHD reported ignoring their disease and engaging in unhealthy behaviors such as smoking, overeating, and/or the use of alcohol.

Anxiety and depression were reported by participants in the present study, and sometimes were hidden from others to avoid "adding another thing for other people to worry about." Uncertainty about future health was one source of anxiety, which is reviewed further in a later section, but other sources of anxiety and sadness included physical limitations and not being able to keep up with peers, school bullying, and other aspects of their life experiences. Cornett & Sims (2014) also reported depression in their participants, along with anxiety related to uncertainty and shame related to physical limitations. This study included adults with a spectrum of CHD, so it is possible that the concerns of adults are different from adolescents and adolescents with SVHD may also have different worries than their counterparts. Anxiety leading to "suffering" was reported by Chiang and colleagues (2015) and related to future uncertainty as well as transition to adult cardiac care. This study included adolescents and young adults with a spectrum of CHD (23% severe CHD). Other quantitative studies have also reported higher levels of anxiety and depression in Fontan survivors (DeMaso et al., 2017; Pike et al., 2018). The mood behaviors identified in this study and others can potentially affect learning and ability for self-care.

I have some limitations, but don't assume: dealing with ableism

The present study as well many previous studies identified physical limitations as a part of life with SVHD survivors, most commonly a varying degree of inability to keep the pace of others during physical exertion or having decreased stamina. McMurray and colleagues (2001) reported adolescent CHD and SVHD survivors described decreased ability to keep up with peers, which sometimes resulted in social isolation and exclusion. Social isolation due to physical

limitations was also noted by Lee and Kim (2012) in their study of adolescents and adults with complex CHD. A range of physical limitations was reported by Shearer and colleagues (2013) in a group of a similar age range, but this study included a spectrum of CHD. In a group of adolescents and adults with SVHD, limitations in physical abilities included poor endurance and sometimes affected social activities, but participants reported being able to find and acknowledge their own tolerated activity pace (Overgaard et al., 2013). These findings were also seen in the present study, with almost all the participants reporting reduced activity endurance of varying degrees, which sometimes resulted in being excluded and social isolation. However, they also found physical activities that they could safely engage in and be successful, such as golf or yoga. In contrast, Chiang et al., (2015) reported that many adolescents and young adults with a spectrum of CHD had negative views of their physical limitations and expressed guilt because of not meeting the academic expectations of their parents due to fatigue symptoms.

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 (Friedman & Owen, 2017). A study by Lee and Kim (2012) also identified that adolescents with complex CHD were subjected to “discriminatory attention” because of their physical limitations and different physical appearance. Adults with complex CHD identified that they had concerns related to the negative effects of their CHD on their employability and insurability (Claessens et al., 2005). Other adolescents with complex CHD and SVHD reported feeling restricted in career choices because of their heart disease (McMurray et al., 2001). Chronic illness and disability are separate concepts that do not automatically coincide, and those who have a chronic illness, such as SVHD, should not be assumed to be disabled, or what their disabilities may be (Wendell, 2001).

One of the more commonly used words by participants in this study, as shown in the word cloud (Figure 5.1) was "condition," rather than "defect" or "disease," demonstrating the use of terminology that does not necessarily indicate a disability.

I see myself as healthy

Another source of disconnect for the adolescents in the present study is that overall, they viewed themselves as generally healthy people. Being labeled as "fragile" or "sick," or assumptions by others of their physical limitations was therefore quite frustrating. Although they acknowledged that they did have some physical limitations, they reported that their heart disease and its limitations did not define them. Others have reported that young adults with SVHD also viewed themselves as healthy (Overgaard et al., 2013), or "healthy with a different shaped heart" (Berghammer et al., 2015).

Adolescence is a time of developing personal identity and one's own body image (Balk, 2014). The participants in the present study expressed that their surgical scars made them feel proud or accomplished or had very little meaning to them. Berghammer et al (2015) also reported that scars were often viewed as a positive affirmation of one's strength and uniqueness in adolescents and adults with SVHD. In another qualitative study from Asia, some participants reported shame or needing to "cover up their scars" (Chiang et al., 2015). Although the patient's scar can be a daily reminder of their heart condition, some adolescents in the present study have perceived their scars in a positive light as an appreciation of health.

I have big plans for the future, but there is also health uncertainty

One of the most important findings of this study, also identified in almost all the previous qualitative studies, was uncertainty related to future health. Despite many quantitatively evaluating their health as the same or somewhat better from last year, ten of 14 (71%)

participants expressed feelings of uncertainty regarding their current or future health status, ranging from not trusting whether their symptoms of activity intolerance were minor enough to "push through," to needing a heart transplant, to an early death. This uncertainty was generally only elicited after asking specifically about it, such as "What do you think your health will be like in the future?" but caused anxiety and fear in the participants, resulting in this subtheme being labeled "the elephant in the room." Other studies have commonly identified uncertainty as a theme in their qualitative findings. The greatest concern among adults with SVHD was death or uncertainty about life expectancy, as well as related concerns about being able to continue working (du Plessis et al., 2018). Anxiety related to uncertainty of illness and possible early death was also reported by in earlier studies. The participants in these studies ranged from adolescents to adults; three studies included only participants with SVHD (Berghammer et al., 2015; du Plessis et al., 2018; Overgaard et al., 2013), three included different types of complex CHD including SVHD (Claessens et al., 2005; Lee & Kim, 2012; McMurray et al., 2001), and the remaining two included a spectrum of mild to complex CHD (Chiang et al., 2015; Cornett & Simms, 2014). Clearly, concerns over possible early death and knowledge of potential lifelong health challenges are common findings and cause anxiety and uncertainty in many SVHD survivors as well as others with CHD.

School can be a challenge for me

One especially problematic area for participants in this study was physical education class, which brought together problems with physical activity limitations and being identified or labeled as different compared to peers. In particular, one challenge that stood out was having to run a mile in physical education class, which is a required standardized task. Some participants reported that their documented activity restrictions were not honored, and they were forced to

participate beyond their ability. Heart disease is often not as visible as other chronic medical conditions, so this may be a reason for teachers not remembering or not honoring their activity limitations. Others reported that exclusion from physical education class was the primary reason that they were labeled as being different.

Differences in physical appearance such as short stature or sternotomy scars, as well as physical activity limitations or need for school accommodations made at least five of the participants in the present study targets for bullying, mostly of the verbal and psychological type. Chiang and colleagues (2015) reported bullying ("being called a freak") in their study of adolescents and young adults with a variety of CHD diagnoses. McMurray (2001) also reported bullying at school in 32% of their participants with severe CHD and SVHD. This was a similar finding in the present study reporting 36% bullying. Bullying occurs in approximately 20% of school students overall ("Bullying Statistics," 2017), and the presence of chronic illness or disabilities increase the risk of bullying (Pittet et al., 2010; Sentenac et al., 2011). School-based bullying prevention programs may decrease bullying by an average of 25% ("Bullying Statistics," 2017).

Participants in this study reported other impacts of SVHD on school success, including difficulty processing information, difficulty focusing on classroom instruction, and other special learning needs. Most of the participants reported some type of school accommodations such as Individualized Education Plans (IEP), extra time for tests or assignments, or attending special education classes. The need for educational accommodations and academic success in adolescents with SVHD is not well studied, one study reported decreased "school competence" or grades and need for educational supports, as well as decreased school QOL in children 8 to 16

years of age with complex CHD, and those with SVHD scored lower than those with other types of CHD (Gerstle et al., 2016).

My quality of life is very good

The participants in this study reported that their quality of life was high, although one participant acknowledged that current physical health concerns negatively impacted HRQOL. One qualitative study on QOL in adolescents with a spectrum of CHD defined QOL as "It's like how you enjoy life" (Shearer et al., 2013). In Shearer's (2013) study, QOL was also reported to be relatively unaffected by CHD. Some participants in the present study also identified that their QOL was perhaps better than people without heart disease because their SVHD made them appreciate life more. This idea was also noted in the qualitative study by Berghammer and colleagues (20015). Furthermore, the quantitative measure of QOL (LAS mean 87.5%) in the present study informs or coincides with the narrative theme identified. However, some of the participants in the present study who were dealing with serious current medical problems reported lower quality of life, related to feeling unwell, stress and anxiety about the potential need for a heart transplant, and not being able to perform activities that were important to them. Because QOL is subjective, it is also possible that because many participants had very little memory of their early surgical experiences, they lacked the frame of reference that remembered early surgeries might add.

One vitally important factor verbalized by participants in dealing with the challenges of SVHD and their uncertain future was the social support from parents and other family members, as well as peers and teachers. One participant stated, "the reason my quality of life is so good is because of the support systems I have." Parents were generally perceived as providing high levels of social support and being treated "normally" helped to shape their perception of being

healthy. Siblings and other family members also provided support and comfort through shared experiences of growing up with SVHD. Friends were a very important source of social support, as expected for the adolescent age group, especially when the participant felt like they were not being treated differently because of their heart disease; this 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 SVHD and other complex CHD (Claessens et al., 2005; Cornett & Simms, 2014; Lee & Kim, 2012; Overgaard et al., 2013; Zahmacioglu et al., 2011).

Another important support identified in this study was heart camp. Many of the participants in this study had previously attended a cardiac-specific, medically supervised summer camp sponsored by a non-profit organization. These types of disease-specific camps provide opportunities for adolescents to share a sense of community and belonging since all the campers share a common medical condition. Potential benefits of these camps include improved self-esteem, self-concept, and QOL 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, and improving disease knowledge and management skills (McCarthy, 2015). Several of the participants in this study had developed long-lasting, deep friendships with fellow campers that continued year-round despite geographical separation.

I have lots of healthcare experiences

Having SVHD means a lifetime of medical care for those affected. The participants in this study often had developed long-lasting relationships with their healthcare team members, which fostered a sense of safety and trust in the adolescents. Previous qualitative studies have reported positive relationships between adolescents and young adults with complex CHD and SVHD and their healthcare providers; these relationships provided a sense of belonging through

emotional support given by the provider (Chiang et al., 2015; Lee & Kim, 2012). Other studies have reported a sense of familiarity and comfort in healthcare experiences (Shearer et al., 2013) and mastery of healthcare experiences (Berghammer et al., 2015). Trust in the healthcare provider is an important factor associated with positive physical and mental health outcomes (Klostermann, Slap, Nebrig, Tivorsak, & Britto, 2005; Robinson, 2016).

In response to the question "What advice would you give doctors and nurses who take care of teens like you?" the current participants tended to focus on being included in the discussion and decision-making, as well as relationship-building and having their questions answered. The participants in the present study were also asked about negative healthcare experiences, which were not reported in previous qualitative studies. Their negative experiences provided opportunities for improvement in communication and perceived caring behaviors.

Implications

Clinical Practice

Adolescents with SVHD face unique challenges which can impact their transition to adulthood. Nurses and other healthcare professionals have many opportunities to positively influence these at-risk youth. Promotion of physical activity appropriate to the individual's SVHD is vital not only for overall cardiac health but overall well-being; rather than an activity restriction. Adolescents with SVHD should be given an activity prescription that is tailored to their unique activity tolerance (Longmuir et al., 2013). This is congruent with the themes of "I am learning to take care of myself" as well as "I have some limitations, but don't assume…" , and "I see myself as healthy". Ongoing developmental assessment to identify evolving or new neurodevelopmental sequelae of SVHD is vital; cognitive screening can be done by outpatient nursing staff quickly during primary or specialty care visits and appropriate referrals should be

made (Pike, Poulsen, & Woo, 2017). Related to the theme of "School can be a challenge", healthcare providers should also ask specifically about the need for school accommodations, school healthcare emergency plans, and inquire specifically about bullying. A new trend in cardiac neurodevelopmental follow-up program is the incorporation of the school liaison role (e.g. typically teacher) to work directly with schools or school districts to communicate educational recommendations from neurodevelopmental testing, act as an advocate for the family and be a liaison between the hospital, family and the school to facilitate obtaining needed services ("School Intervention Program | Children's Hospital of Wisconsin," 2019). This model has been implemented very successfully at several cardiac neurodevelopmental programs and although it adds cost, may be a wise investment of resources. School nurses can provide vital support to enacting school accommodations in place without resulting in social isolation for the adolescent with SVHD, as well as helping teachers understand the student's physical limitations. In addition, school nurses should be aware of any emergency medical needs such as arrhythmias or presence of a pacemaker or internal defibrillator for an adolescent student with SVHD and collaborate with specialty care providers to form an appropriate emergency care plan.

Healthcare providers should be sensitive to the developmental, cultural, and privacy needs of the adolescent with SVHD, and involve them in healthcare-related decision-making and problem-solving. Nurses can assist adolescents with SVHD to be an active participant in healthcare visits and to advocate for the adolescent's autonomy. These interventions will assist the adolescent with SVHD to learn self-care. Anxiety and depression should be anticipated, screened for, discussed and appropriate referrals made. Uncertainty is common, often with resulting anxiety; this should also be anticipated and acknowledged while providing the adolescent with honest information about their current health and anticipated future health.

Adolescents with worsening physical health status should be assisted to identify support and resources to help them cope. Healthcare providers should continue to provide information and referrals to heart camps to allow adolescents with SVHD to meet others like themselves, developing relationships with like-peers, and learn new coping strategies through these supportive relationships. There are many support groups for parents and families of children with CHD, included Mended Little Hearts and California Heart Connect, but the purpose of these groups is primarily to support families and to provide resources to help their child. The available support groups are not designed to specifically support the adolescent with SVHD or other complex CHD. Given the small numbers of affected adolescents in all but the largest communities, it may be necessary to combine a SVHD support group for adolescents with young adults or to move to an online format. There are many online groups and websites providing information, support, and advocacy; a directory of these resources could be beneficial for the adolescent seeking to gain information or join an online group.

Transition to adult cardiac care begins during adolescence. The American Heart Association has recently published best practices for the comprehensive transition of adolescents and young adults with CHD, including those with SVHD (Sable et al., 2011). This Scientific Statement recommends that transition include the adolescent and the parents and incorporate a "medical home" for primary and specialty care. Based on the findings in this study, it may be beneficial to begin the transition interventions and support earlier, since many of the participants had not started the process by the age of 17 years. Recommended transition program components include cardiac health and follow-up, as well as genetic counseling, guidance on reproductive health and family planning, exercise prescription, education, and employment, obtaining insurance, psychological health, QOL, mortality and advance directives (Sable et al., 2011).

These programs utilize nurses as well as advanced practice nurses, as well as other disciplines. Transition programs should consider the need for psychological support and the beneficial influence of peers; a peer-led support group with input and guidance of a multi-disciplinary team of nursing, psychology or social work might be helpful. Some of the heart camps also offer a transition program that includes weekend trips to assist young adults with transition-related planning such as obtaining health insurance, employability, and living skills (https://www.campdelcorazon.org/portal/p.a.c.e.).

Research

This study has provided insights into the daily lives of adolescents with SVHD, specifically related to school experiences, relationships with friends and family, and perceptions of their healthcare experiences. The findings from this study are limited in generalizability due to its qualitative nature but are also quite congruent with the findings of other qualitative studies of Fontan survivors from other parts of the world. The goal of phenomenological research is to better understand the experience being studied in order to help generate further research questions or to guide future studies of interventions. The unique challenges that emerged in this study included school-related issues related to physical challenges (often accommodations not acknowledged or respected) and health, absenteeism, bullying, being identified as different, and being labeled warrants additional investigations using both quantitative and qualitative methods. Adding the perspectives of teachers, school nurses, and parents would provide valuable information. Strategies or programs that may support success in school should be evaluated. In addition, the effects of transition programs from pediatric to adult cardiac care must be quantified. Further research is also needed on effective and culturally sensitive adolescent support strategies and how best to support families with children affect by heart disease.

Health Policy

Dr. Jack Rychik, a well-known pediatric cardiologist and a leading researcher on Fontan physiology, recently stated at a conference, "We created these unique [Fontan] survivors, we, therefore, have an obligation to them" (personal communication, February 13, 2019). Dr. Rychik was referring to the need for evidence-based, innovative medical management strategies for the life-limiting sequelae of this unique cardiac physiology (Rychik, 2016). However, his words also evoke a sense of responsibility for healthcare providers as well as the community and society, to support these unique survivors and their families. Access to healthcare and insurance, mental healthcare, family medical leave to care for a child with a serious health condition and funding for expanded school services are just some of the important healthcare policy issues that are relevant. School liaison programs to bridge the cardiac neurodevelopmental program recommendations to the school system provide advocacy and support for children and adolescents with SVHD and their families, but these types of programs need financial support. Programs that support the transition from pediatric to adult healthcare provide important services and funding for these programs should be supported. After transitioning to adulthood, Fontan survivors must also deal with insurability with a pre-existing condition, need for further education or employment protections and accommodations, access to healthcare including mental health care, disability insurance, and long-term care.

Limitations and Conclusions

Limitations

Purposive sampling has inherent selection bias but is commonly used in qualitative research and was necessary because the population of adolescents with SVHD is small, and the use of other sampling methods would render the study not feasible. The adolescents who agreed to

participate in the study were not representative of all Fontan survivors; most potential participants were approached because they had previously participated in research and were known to be able to describe their experiences well. The participants likely over-represent Fontan survivors who were medically relatively well, because adolescents who were currently listed for organ transplant were excluded and those with severe neurodevelopmental and psychological co-morbidities were not approached to participate. The experiences of Fontan survivors who chose not to participate in research or who were not approached may be different. Phenomenological research requires interpretation by the researchers, which may lead to researcher bias. The impact of the researchers' experience with this population was carefully considered and strategies to limit the effect of reflexivity were employed. Appropriate strategies to support the trustworthiness of the research findings were implemented and described in the methods.

The phenomenological approach requires that participants be articulate in describing their experiences, which may be difficult if the participant experiences cognitive or communicative difficulties as the result of their SVHD. These difficulties may not be apparent until the interview, which may then have required including additional participants. It is possible that some cognitive or neurodevelopmental difficulties were not apparent to the researchers but influenced participants' responses. However, the purpose of phenomenological research is to understand an experience as the participant perceives and lives it, regardless of how that experience is influenced. It is also assumed that research participants were truthful and forthcoming during their interview. Phenomenological research is not generalizable but does yield highly valuable understanding into lived experiences and may help guide theory development.

Conclusion

Adolescents with SVHD are thriving in the face of uncertainty. They are pioneers in many ways and are the first generation to reliably survive to adulthood with highly complex heart disease. They face many physical, psychosocial and academic challenges, as well as an uncertain future. Despite these challenges, they perceive themselves as healthy individuals with a high QOL, and many perceive that having SVHD contributes to their uniqueness as individuals and allows them to more fully appreciate their lives. They are generally able to adapt to their activity limitations and find ways to succeed in the context of their physical abilities. However, they also experience anxiety and sadness related to uncertainty about their future. Their support systems in the ecological niches of family, peers, school and social media provide them with resources to cope with the challenges associated with SVHD. The results of this study support findings of previous qualitative research in this population and add further information on experiences of school, social relationships, and healthcare experiences not previously reported in this population of unique adolescents.

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Appendix A. Recruitment Flyer

Experiences of Teen and Young Adults with Single Ventricle Heart Disease

UCLA Research Study

Participants Needed

Are you 14-21 years of age with a single ventricle heart disease and have undergone the Fontan procedure?

The purpose of this study is to gain understanding of the everyday experiences of school, relationships, and healthcare encounters in teens and young adults who have single ventricle heart disease and have had a Fontan operation in the past. Participants will be asked to complete a demographic and clinical information form (20 minutes), and participate in one in-person interview on the UCLA campus to talk about your experiences (about 1 hour). Participation is voluntary.

Participants will receive $50 for participation in the study. For more information, please contact Dr. Nancy Pike at 310-903-2614 or email npike@sonnet.ucla.edu

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Appendix B. Recruitment Script.

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

SCRIPT FOR INITIAL SCREENING THROUGH THE TELEPHONE

The following script would be used in potential subjects who participated in Dr. Pike's brain MRI study and checked the box on the consent form indicating that they were willing to be contacted regarding future research studies. They will be speaking with a research assistant from UCLA.

The bracketed text would be used only if a potential participants had asked for think about or discuss participation during the recruitment call, otherwise, the recruitment call would continue to screening {Hello, this is [name of caller] from UCLA School of Nursing. You/your child previously participated in Dr. Nancy Pike's brain MRI study and indicated that you were interested in participating in a new study, titled "School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease".
This research study is being conducted by Principal Investigator, Dr. Nancy Pike and Co-Investigator Jennifer Peterson. If you have a few moments, I would like to tell you a little bit about the research.

The purpose of the study is to better understand how having single ventricle heart disease affects the everyday experiences of ten and young adults. These everyday experiences include school, relationships with teachers, family, and friends, as well as healthcare experiences. It is very important that healthcare providers understand how having single ventricle heart disease affects people so that they can provide better care. If you or your child are eligible, your participation in the research will involve filling out a demographic and clinical information form, and participating in an interview.}

I need to ask you a few questions in order to determine whether you/your child may be eligible for the research. I will ask you about you/your child's age and recent medical history. Would you like to continue with the screening? The screening will take about 5 minutes or less. You do not have to answer any medical history questions that may make you feel uncomfortable and can choose to stop at any time. Your participation in the screening is voluntary and is of no direct benefit to you. A decision whether or not to participate in the screening will not affect your relationship with UCLA.

Your answers will be confidential. No one will know your answers except for the research team. Once we are through with the screening interview, I will determine whether or not you are eligible for the study based on your answers. If you are eligible, we will keep this information and it will be kept in a file without your name or other identifying information. If you are eligible and decide to participate, you will also sign an informed consent form on the date of study participation. This will also be kept in our research records and will have your name on it. However, if you are not eligible, all information you provided to me during this screening will be destroyed.
Would you like to continue with the screening?

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[*If no, thank the person and hang-up*]. [*If yes, continue with the screening*].

For Congenital Heart Disease Group:
Do you or your child have a single ventricle heart defect? (Yes / NO) Have you

or your child undergone surgical repair or palliation? (Yes / NO)

Have you or your child had a Fontan operation, or final stage of surgery for single ventricle heart disease? (Yes/NO)

Are you or your child, 14 –21 years old? (Yes / NO)

Do you or your child have severe developmental delay that would make filling out a questionnaire or participating in a verbal interview not possible? (Yes/NO)

Have you or your child had an organ transplant? (Yes/NO)

Are you, or your child currently listed for an organ transplant? (Yes/NO) [*If*

yes, include the following at the end of the screening]:
Thank you for answering the screening questions.
[Indicate whether the person is eligible, requires additional screening at the clinic, or is not eligible and explain why.]

If the potential subject does <u>not</u> meet the study criteria
Because you responded "yes" that you have *[developmental delay, organ transplant, or listed for organ transplant – state which one]* or you responded "no" that you are *[not had a Fontan operation, single ventricle heart disease, or are outside the age range of 14 to 21 years of age- state which one],* you are NOT eligible for the study. *[Thank the person and hang-up]*

If the potential subject meets the study criteria
I have a copy of the consent that I can mail/email to you or give you in person at your next clinic appointment for your review. I can also read the consent to you now over the telephone. Please feel free to let us know if you have any questions that we can answer for you. Just to clarify, if you choose to participate in the study the consent form will be signed at your in-person visit before any study procedures because emailing, mailing, or reading the consent over the phone does not constitute the consent process.

If you agree to participate, you and your child will sign the consent / assent form at the time of your appointment. At your appointment, you will be asked to complete a demographic and clinical information form and participate in an interview that will take a total of approximately 2 hours of your time. You will be reimbursed $50.00 dollars cash for participating in the study.

This study is not being done to improve your condition or health. The primary risks for the study procedures include feelings of nervousness about completing the study measures. It is also possible that some of the interview questions could cause you to remember times when you felt sad or afraid. You can pause or stop the interview at any time, and do not have to answer questions that you don't want to answer. Your participation is voluntary. If you chose not to participate, that will not affect your right to health care or other services to which you are otherwise entitled. All information you provide will be kept confidential.

Do you have any questions about the screening or the research? I am going to give you a couple of telephone numbers to call if you have any questions later. Do you have a pen? If you have questions about the research screening, you may call one of our Principal Investigators:
Dr. Nancy Pike at 310-903-2614 (cell) or 310-206-3683 (work) or
Jennifer Peterson at 714-280-3219 (cell) or 562-933-3308 (work).

If you have questions regarding the rights of research subjects or if you have complaints or concerns about the research and cannot reach the Principal Investigator; or just want to talk to someone other than the Investigator, you may call the UCLA Office of the Human Research Protection Program at (310) 825-7122.

Thank you for your willingness to answer our questions. Have a nice day.

Name: _____
Address: _____
E-mail: _____
Phone Number: _____

Screener's Name (print) _____

Screener's Signature _____

Screen Date: _____

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Ventrículo Único

GUIÓN PARA LA SELECCIÓN INICIAL A TRAVÉS DEL TELÉFONO

El siguiente guión se usaría en posibles sujetos que participaron en el estudio de resonancia magnética cerebral de la Dra. Pike y marcaron la casilla en el formulario de consentimiento que indica que estaban dispuestos a ser contactados con respecto a futuros estudios de investigación. Ellos estarán hablando con un asistente de investigación de UCLA.

El texto entre corchetes se usaría solo si los participantes potenciales hubieran pedido Tiempo para pensar o discutir la participación durante la llamada de selección, de lo contrario, la llamada de selección continuaría. {Hola, le llama [nombre de la persona que llama] de UCLA Escuela de Enfermería. Usted / su hijo participó anteriormente en el estudio de resonancia magnética cerebral de la Dra. Nancy Pike e indicó que estaba interesado en participar en un nuevo estudio, titulado "Escuela y Experiencias de Vida de Adolescentes y Adultos Jóvenes con Enfermedad Cardíaca de Ventrículo Único". Este estudio de investigación está siendo llevado a cabo por la investigadora principal, la Dra. Nancy Pike y Co-Investigadora Jennifer Peterson. Si tiene unos momentos, me gustaría contarles un poco sobre la investigación.

El propósito del estudio es comprender mejor cómo la enfermedad cardiaca de un solo ventrículo afecta las experiencias cotidianas de adolescentes y adultos jóvenes. Estas experiencias diarias incluyen la escuela, las relaciones con los maestros, la familia y los amigos, así como experiencias de salud. Es muy importante que los proveedores de atención médica entiendan cómo el tener enfermedad cardíaca de un solo ventrículo afecta a las personas para que puedan brindar una mejor atención. Si usted o su hijo son elegibles, su participación en la investigación implicará completar un formulario de información demográfica y clínica, y participación en una entrevista.}

Necesito hacerle algunas preguntas para determinar si usted o su hijo pueden ser elegible para la investigación. Le preguntaré acerca de la edad de usted / su hijo y de su historial médico. ¿Le gustaría continuar con el proceso de selección? El proceso tomará alrededor de 5 minutos o menos. No tiene que contestar ninguna pregunta sobre su historial médico que pueda hacerle sentir incómodo y puede optar por terminar en cualquier momento. Su participación en el este proceso es voluntaria y no le beneficia directamente. Una decisión de si o no participar en el proceso de selección no afectará su relación con UCLA.

Sus respuestas serán confidenciales. Nadie sabrá sus respuestas, excepto el equipo de investigación. Una vez que hayamos terminado con la entrevista de selección, determinaré Si es o no elegible para el estudio basado en sus respuestas. Si es elegible, mantendremos esta información y la guardaremos en un archivo sin su nombre ni información identificativa. Si es elegible y decide participar, también firmará un formulario de consentimiento informado en la fecha de participación en el estudio. Esto también se mantendrá en nuestros registros y tendrá su nombre en él. Sin embargo, si no es elegible, toda la información que me proporcionó durante esta evaluación será destruida.

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[Si no, agradezca a la persona y cuelgue]. [Si es así, continúe con la entrevista].

Para el grupo de enfermedad cardíaca congénita:

¿Usted o su hijo tienen defecto cardíaco de un solo ventrículo? (Si / No)

¿Usted o su hijo han sido sometidos a reparación quirúrgica o paliación? (Si / No)

¿Ha tenido usted o su hijo una operación de Fontan, o la etapa final de la cirugía para cardiopatía de ventrículo único? (Si / No)

¿Es usted o su hijo de 14 a 21 años? (Si / No)

¿Usted o su hijo tienen un retraso grave en el desarrollo que haría completar un cuestionario o participación en una entrevista verbal imposible? (Si / No)

¿Ha tenido usted o su hijo un trasplante de órganos? (Si / No)

¿Está usted o su hijo actualmente en la lista para un trasplante de órganos? (Si / No)

[En caso afirmativo, incluya lo siguiente al final de la evaluación]:
Gracias por responder a las preguntas de selección.
[Indique si la persona es elegible, requiere una evaluación adicional en la clínica o si no es elegible y explique por qué.]

Si el sujeto potencial no cumple con los criterios del estudio.
Debido a que respondió que "sí" tiene [retraso en el desarrollo, trasplante de órganos o listado para trasplante de órganos - indique cuál] o porque respondió que "no" [tuvo una operación de Fontan, tiene una enfermedad cardíaca de ventrículo único o está fuera del rango de edad de de 14 a 21 años de edad, indique cuál de ellos], NO es elegible para el estudio. *[Agradezca a la persona y cuelgue]*.

Si el sujeto potencial cumple con los criterios de estudio.
Tengo una copia del consentimiento que puedo enviarle por correo / correo electrónico o entregarle personalmente en su próxima cita clínica para su revisión. También puedo leer el consentimiento a usted ahora sobre el teléfono. Por favor, no dude en hacernos saber si tiene alguna pregunta que podamos responderle. Solo para aclarar, si decide participar en el estudio, el formulario de consentimiento se firmará en su visita en persona antes de cualquier procedimiento de estudio debido a que el envío de correos electrónicos, el envío por correo o la lectura del consentimiento por teléfono no constituyen el proceso de consentimiento.

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hora de su cita. En su cita, se le pedirá que complete unos formularios de información demográfica y clínica y que participe en una entrevista y tardara una total de aproximadamente 2 horas de su tiempo. Se le reembolsará $ 50.00 dólares en efectivo por participar en el estudio.

Este estudio no se está haciendo para mejorar su condición o salud. El riesgo principal para los procedimientos del estudio incluye sentimientos de nerviosismo al completar el estudio.
También es posible que algunas de las preguntas de la entrevista puedan hacer que recuerde los momentos en que se sintió triste o con miedo. Puede pausar o detener la entrevista en cualquier momento, y no tiene que responder preguntas que no desea responder. Su participación es voluntaria. Si elige no participar, eso no afectará su derecho a atención médica u otros servicios a los que tiene derecho. Toda la información que proporcioné se mantendrá confidencial.

¿Tiene alguna pregunta sobre la entrevista o la investigación? Voy a darle un par de números de teléfono para llamar si tiene alguna pregunta más adelante. ¿Tiene un bolígrafo? Si tiene preguntas sobre la investigación de detección, puede llamar a uno de nuestros
Investigadores principales:

Dr. Nancy Pike al 310-903-2614 (celular) o 310-206-3683 (trabajo) o

Jennifer Peterson al 714-280-3219 (celular) o 562-933-3308 (trabajo).

Si tiene preguntas sobre los derechos de los sujetos de investigación o si tiene quejas
o inquietudes sobre la investigación y no puede comunicarse con el Investigador Principal; o si desea hablar con alguien que no sea el Investigador, puede llamar a la Oficina de UCLA del Programa de Protección de Investigación Humana al (310) 825-7122.

Gracias por estar dispuesto a responder nuestras preguntas. Que tengas un buen día.

Nombre: _____
Dirección:_____
Email:_____
Número de teléfono: _____
Nombre del investigador (en letra de imprenta) _____
Firma del investigador _____
Fecha de la pantalla: _____

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Appendix D. Informed Consent

UNIVERSITY OF CALIFORNIA LOS ANGELES
CONSENT TO PARTICIPATE IN RESEARCH

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

INTRODUCTION

You are invited to participate in a research study conducted by Dr. Nancy Pike, PhD from the University of California, Los Angeles (UCLA) School of Nursing, and Jennifer Peterson PhD(c) from the School of Nursing at University of California, Irvine. You are invited to participate in this study because you are a young adult between the ages of 18 and 21 years with single ventricle heart disease and have had a Fontan operation in the past. Participation in this study is completely voluntary. Please read the information below, and ask questions about anything you do not understand, before deciding whether or not to participate.

PURPOSE OF THE STUDY?

The purpose of this study is to better understand the impact of single ventricle heart disease on daily life, including school experiences, relationships with family and friends, and healthcare experiences. As treatments for congenital heart disease have improved over the past several decades, more and more children with single ventricle heart disease have survived to adulthood. Some aspects of having single ventricle heart disease, such as the effects of open heart surgery, being cyanotic (having lower blood oxygen levels), having lower activity tolerance or having abnormal heart rhythms may affect people's lives in many ways. Some of these effects might include how teens and young adults with single ventricle heart disease experience school as well as relationships with family and friends. In addition, people with single ventricle heart disease need life-long medical care and their previous healthcare experiences may influence how they view their current and future healthcare experiences. To provide the best possible care, it is important for healthcare providers to understand how teens and young adults with single ventricle heart disease see their lives and experiences.

WHAT WILL HAPPEN IF I TAKE PART IN THIS STUDY?

If you volunteer to participate in this study, we would ask you to do the following things:
Before you begin the study:

1. Work with the researchers to schedule an in-person visit to complete the study measures, including an interview and a written survey.

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2. Provide verbal and written informed consent for your study participation on the day of the interview. The interview will take place at the UCLA campus, or your home if preferred.

During the study:

1. Sign a separate Health Insurance Portability and Accountability (HIPAA) research authorization form to allow researchers to review your medical records regarding your heart diagnosis and history, type and dates of surgeries, oxygen saturation level, medications, height and weight and other medical conditions.
2. Complete a demographic and clinical information form about your living situation, school, family structure, and current medical status (about 20 minutes).
3. Participate in an interview with the researcher to talk about your experiences in school, relationships with family and friends, and healthcare experiences. The interview will be audio recorded (about 60 minutes).
4. If the researchers have additional questions that arise after the interview, they may contact you to set up an additional voluntary interview, which may be done in-person or by phone.

HOW LONG WILL I BE IN THIS STUDY?

This study will last about 1 ½ to 2 hours on the study day. If a second interview is requested, it will occur within 3 to 5 months of the first interview.

WHAT KINDS OF RISKS OR DISCOMFORTS COULD I EXPECT?

Known risks and discomforts:

The possible risks and/or discomforts associated with the procedures described in this consent form include:

- There may be some nervousness about participating in the interview.
- There is a very small risk that talking during the interview might make you remember times that you felt sad, scared, or had your feelings hurt, and although this is rare, it may cause you to feel uncomfortable. You do not have to answer any question that makes you uncomfortable. If you want to stop during the interview, either for a few minutes or stop completely, you can do so.
- There is a very small risk that your confidentiality could be broken. There are many measures in place to prevent that from happening, described on page 3 and page 4.

Unknown risks and discomforts:
The researchers will let you know if they learn anything that might make you change your mind about participating in the study.

ARE THERE ANY BENEFITS IF I PARTICIPATE?

Possible benefits to me:
You will not directly benefit from your participation in this research.

Possible benefits to others or society:
This study will help the researchers learn more about living with complex congenital heart disease and how this type of heart disease impacts daily activities. This information may help in the treatment of future patients with single ventricle heart disease who have had a Fontan procedure.

WHAT OTHER CHOICES DO I HAVE IF I DON'T WANT TO PARTICIPATE?

If you decide not to take part in this study, or if you withdraw from this study before it is completed, your medical care and relationships with your healthcare providers will not change in any way.

PARTICIPATION AND WITHDRAWAL

Your participation in this research is voluntary. Your choice about whether or not to participate will have no effect on your care, services, or benefits at UCLA. If you agree to participate, but later decide to withdraw from this study, you may do so without affecting your rights to care, services, or benefits at UCLA. If you decide to stop being in the study, the researcher will ask you to let them know as soon as possible. You may review, edit or erase the audiotape of your interview at your request. If you decide to withdraw from the study, your audiotapes and transcribed interview will be destroyed. If you decide to edit the interview audiotape, the researcher may ask you to do so in another audiotaped interview.

CAN THE RESEARCHERS REMOVE ME FROM THIS STUDY?

The researchers may end your participation in this study if necessary to protect your health or if other situations arise that make it necessary to do so (such as continued visible distress during the interview).

HOW WILL INFORMATION ABOUT ME AND MY PARTICIPATION BE KEPT CONFIDENTIAL?

The researchers will do their best to make sure that your private information is kept confidential. Information about you will be handled as confidentially as possible but participating in research may involve a loss of privacy and the potential for a breach in confidentiality. Study data will be physically and electronically secured. As with any use of electronic means to store data, there is a risk of breach of data security.

Under California law, the researchers cannot keep information about known or reasonably suspected incidents of abuse or neglect of a child, dependent adult, or elder confidential and must report to authorities. This includes physical, sexual, emotional, or financial abuse or neglect.

The researchers may not be able to keep confidential any disclosure of thoughts to harm yourself. If you disclose thoughts of suicide or harming yourself, you will be referred to the closest emergency room for further assessment and evaluation.

Use of personal information that can identify you:
Any information that is obtained in connection with this study and that can identify you will remain confidential. It will be disclosed only with your permission or as required by law. Confidentiality will be maintained by assigning you a unique study identification number that is used to identify your written and audiotaped responses. The key to the study identification numbers is kept in a locked file cabinet, separate from the study data.
How information about you will be stored:
The study data and records are kept in a locked file cabinet in the investigator's office. Electronic data, including transcribed interviews, will be kept on an encrypted, password protected computer drive.

People and agencies that will have access to your information:
Only the research team will have access to the study data. The research team and authorized UCLA personnel may have access to study data and records to monitor the study. Research records provided to authorized, non-UCLA personnel will not contain identifiable information about you. Publications and/or presentations that result from this study will not identify you by name.

How long information from the study will be kept:
Information from the study, including the code list of identifiers and questionnaires, will be kept for three years after study completion, then destroyed.

WILL I BE PAID FOR MY PARTICIPATION?

You will be given $50 for participation in this study on the study day to compensate you for travel and parking costs as well as your time and inconvenience of participating in the study. You will be asked to sign a receipt of payment form.

WHO CAN I CONTACT IF I HAVE QUESTIONS ABOUT THIS STUDY?

The Research Team:
You may contact Dr. Nancy Pike at 310-903-2614; npike@sonnet.ucla.edu, or Jennifer Peterson at 714-280-3219; jkpeter1@uci.edu with any questions or concerns about the research or your participation in this study.

UCLA Office of the Human Research Protection Program (OHRPP):
If you have questions about your rights while taking part in this study, or you have concerns or suggestions and you want to talk to someone other than the researchers about the study, you may contact the UCLA OHRPP by phone: (310) 206-2040; by email: participants@research.ucla.edu or mail: Box 951406, Los Angeles, CA 90095- 1406.

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WHAT ARE MY RIGHTS IF I TAKE PART IN THIS STUDY?
Taking part in this study is your choice. You can choose whether or not you want to participate. Whatever decision you make, there will be no penalty to you and you will not lose any of your regular benefits.

- You have a right to have all of your questions answered before deciding whether to take part.
- Your decision will not affect the medical care you receive from UCLA.
- If you decide to take part, you can leave the study at anytime.
- If you decide to stop being in this study, you should notify the research team right away. The researchers may ask you to complete some procedures in order to protect your safety.
- If you decide not to take part, you can still get medical care from UCLA.

Contact for Future Research

The researchers may contact me in the future to ask me to take part in other single ventricle heart disease research studies.

☐ YES NO

HOW DO I INDICATE MY AGREEMENT TO PARTICIPATE?
If you agree to participate in this study, you should sign and date below.

Your signature below indicates:

- You have read this document and understand its meaning
- You have had a chance to ask questions and have had these questions answered to your satisfaction
- Your consent to your participation in this research study; and
- You will be given a signed copy of this form and a signed copy of the HIPAA authorization form.

SIGNATURE OF THE PARTICIPANT

_____
Name of Participant

_____ _____
Signature of Participant Date

SIGNATURE OF PERSON OBTAINING CONSENT
I have explained the research to the participant and/or the participant's parent(s)/legal guardian(s) and have answered all of their questions. I believe that they understand all of the information described in this document and freely give consent to participate.

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Name of Person Obtaining Consent	Contact Number
Signature of Person Obtaining Consent	Date

Routing of signed copies of the form: 1. Give to adult subject or parent/legal guardian (copy). 2. Place in Principal Investigator's research file (original)

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Appendix E. Parent Permission University of California, Los Angeles
PARENT PERMISSION FOR MINOR TO PARTICIPATE IN RESEARCH

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

Dr. Nancy Pike, PhD from the University of California, Los Angeles (UCLA) School of Nursing, and Jennifer Peterson PhD(c) from the School of Nursing at University of California are conducting a research study.

Your child was selected as a possible participant in this study because he/she is between the ages of 14 and 17 years with single ventricle heart disease and has had a Fontan operation in the past. Your child's participation in this research study is voluntary.

Why is this study being done?

The purpose of this study is to better understand the impact of single ventricle heart disease on daily life, including school experiences, relationships with family and friends, and healthcare experiences. As treatments for congenital heart disease have improved over the past several decades, more and more children with single ventricle heart disease have survived to adulthood. Some aspects of having single ventricle heart disease, such as the effects of open heart surgery, being cyanotic (having lower blood oxygen levels), having lower activity tolerance or having abnormal heart rhythms may affect people's lives in many ways. Some of these effects might include how teens and young adults with single ventricle heart disease experience school as well as relationships with family and friends. In addition, people with single ventricle heart disease need life-long medical care and their previous healthcare experiences may influence how they view their current and future healthcare experiences. To provide the best possible care, it is important for healthcare providers to understand how teens and young adults with single ventricle heart disease see their lives and experiences.

What will happen if my child takes part in this research study?

If you agree to allow your child to participate in this study, we would ask him/her to:

Before the study:

1. Work with the researchers to schedule an in-person visit to complete the study measures, including an interview and a written survey.
2. Provide verbal and written informed assent for study participation on the day of the interview. The interview will take place at the UCLA campus, or your home if preferred.

During the study:

1. The parent or guardian will be asked to sign a separate Health Insurance Portability and Accountability (HIPAA) research authorization form to allow

researchers to review your child's medical records regarding your heart diagnosis, type and dates of surgeries, oxygen saturation level, medications and other medical conditions.
2. Complete a demographic and clinical information form about their living situation, school, family structure, and current medical status (about 20 minutes).
3. Participate in an interview with the researcher to talk about their experiences in school, relationships with family and friends, and healthcare experiences. The interview will be audio recorded (about 60 minutes).
4. If the researchers have additional questions that arise after the interview, they may contact your child to set up an additional voluntary interview, which may be done in-person or by phone.

How long will my child be in the research study?

Participation will take a total about 1 ½ to 2 hours on the study day. If a second interview is requested, it will occur within 3 to 5 months of the first interview.

Are there any potential risks or discomforts that my child can expect from this study?
The possible risks and/or discomforts associated with the procedures described in this consent form include:

- There may be some nervousness about participating in the interview.
- There is a very small risk that talking during the interview might make your child remember times that they felt sad, scared, or had their feelings hurt, and although this is rare, it may cause your child to feel uncomfortable. Your child does not have to answer any question that makes them uncomfortable. If your child wants to stop during the interview, either for a few minutes or stop completely, they can do so.
- There is a very small risk that your child's confidentiality could be broken. There are many measures in place to prevent that from happening, described on page 3 and page 4.

Unknown risks and discomforts:
The researchers will let you and your child know if they learn anything that might make you change your mind about participating in the study.

Are there any potential benefits to my child if he or she participates?

Your child will not directly benefit from your participation in the research.
The results of the research may help the researchers learn more about living with complex congenital heart disease and how this type of heart disease impacts daily activities. This information may help in the treatment of future patients with single ventricle heart disease who have had a Fontan procedure.

Will my child be paid for participating?
Your child will receive $50 for participation in this study on the study day to compensate for travel and parking costs as well as time and inconvenience of participating in the study. Your child will be asked to sign a receipt of payment form.

Will information about my child's participation be kept confidential?

The researchers will do their best to make sure that your child's private information is kept confidential. Information about your child will be handled as confidentially as possible but participating in research may involve a loss of privacy and the potential for a breach in confidentiality. Study data will be physically and electronically secured. As with any use of electronic means to store data, there is a risk of breach of data security.

Under California law, the researchers cannot keep information about known or reasonably suspected incidents of abuse or neglect of a child, dependent adult, or elder confidential and must report to authorities. This includes physical, sexual, emotional, or financial abuse or neglect.

The researchers may not be able to keep confidential any disclosure of your child's thoughts to harm themselves. If your child discloses thoughts of suicide or self-harm yourself, you and your child will be referred to the closest emergency room for further assessment and evaluation.

Use of personal information that can identify you:
Any information that is obtained in connection with this study and that can identify your child will remain confidential. It will be disclosed only with your permission or as required by law. Confidentiality will be maintained by assigning your child a unique study identification number that is used to identify written and audiotaped responses. The key to the study identification numbers is kept in a locked file cabinet, separate from the study data.

How information about you will be stored:
The study data and records are kept in a locked file cabinet in the investigator's office. Electronic data, including transcribed interviews, will be kept on an encrypted, password protected computer drive.

People and agencies that will have access to your information:

Only the research team will have access to the study data.

The research team and authorized UCLA personnel may have access to study data and records to monitor the study. Research records provided to authorized, non-UCLA personnel will not contain identifiable information about your child. Publications and/or presentations that result from this study will not identify your child by name.

How long information from the study will be kept:
Information from the study, including the code list of identifiers and questionnaires, will be kept for three years after study completion, then destroyed.

- **Withdrawal of participation by the investigator**

The investigator may withdraw your child from participating in this research if circumstances arise which warrant doing so. If necessary to protect your child's health, such as continued visible distress during the interview, your child may have to drop out, even if they would like to continue. The investigator will make the decision and let your child know if it is not possible for him/her to continue. The decision may be made to protect your child's health and safety.

What are my and my child's rights if he or she takes part in this study?

- You can choose whether or not you want your child to be in this study, and you may withdraw your permission and discontinue your child's participation at any time.
- Whatever decision you make, there will be no penalty to you or your child, and no loss of benefits to which you or your child were otherwise entitled.
- Your child may refuse to answer any questions that he/she does not want to answer and still remain in the study. Your child may review, edit or erase the audiotape of your interview at your request. If you or your child decide to withdraw from the study, your child's audiotapes and transcribed interview will be destroyed. If your child decides to edit the interview audiotape, the researcher may ask them to do so in another audiotaped interview.

Who can I contact if I have questions about this study?

- **The research team:**

If you have any questions, comments or concerns about the research, you can talk to the one of the researchers. Please contact Dr. Nancy Pike at 310-903-2614; npike@sonnet.ucla.edu, or Jennifer Peterson at 714-280-3219; jkpeter1@uci.edu with any questions or concerns about the research or your child's participation in this study.

- **UCLA Office of the Human Research Protection Program (OHRPP):**
  If you have questions about your rights as a research subject, or you have concerns or suggestions and you want to talk to someone other than the researchers, you may contact the UCLA OHRPP by phone: (310) 206-2040; by email: participants@research.ucla.edu or by mail: Box 951406, Los Angeles, CA 90095- 1406.

Contact for Future Research

The researchers may contact me in the future to ask me to take part in other single ventricle heart disease research studies.

☐ YES NO

You will be given a copy of this information to keep for your records.

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Committee: South General IRB

SIGNATURE OF PARENT OR LEGAL GUARDIAN

Name of Child

Name of Parent or Legal Guardian

Signature of Parent or Legal Guardian | Date

SIGNATURE OF PERSON OBTAINING CONSENT

Name of Person Obtaining Consent | Contact Number

Signature of Person Obtaining Consent | Date

Protocol ID:IRB#18-001204 UCLA IRB Approved Approval Date: 12/2/2018 Through: 9/19/2021
Committee: South General IRB

Appendix F. Parent Permission Spanish
University of California, Los Angeles

PERMISO DE PADRES POR MENOR PARA PARTICIPAR EN LA INVESTIGACIÓN

Escuela y Experiencias de Vida de Adolescentes y Adultos Jóvenes con Enfermedad Cardíaca de Ventrículo Único

Dr. Nancy Pike, PhD de la Facultad de La Escuela de Enfermería de la Universidad de California, Los Ángeles (UCLA), y Jennifer Peterson PhD(c) de la Escuela de Enfermería de la Universidad de California están llevando a cabo un estudio de investigación.

Su hijo fue seleccionado como posible participante en este estudio porque tiene entre 14 y 17 años, tiene enfermedad cardíaca de ventrículo único y ha tenido una operación de Fontan en el pasado. La participación de su hijo en este estudio de investigación es voluntaria.

¿Por qué se está haciendo este estudio?
El propósito de este estudio es comprender mejor el impacto de la enfermedad cardíaca de ventrículo único en la vida diaria, incluyendo las experiencias escolares, las relaciones con familiares y amigos, y experiencias médicas. Como tratamientos para la cardiopatía congénita han mejorado en las últimas décadas, cada vez más niños con enfermedad cardíaca de ventrículo único han sobrevivido hasta la edad adulta. Algunos aspectos de tener un solo ventrículo cardíaco, como los efectos de la cirugía a corazón abierto, ser cianótico (tener bajos niveles de oxígeno en la sangre), tener menor tolerancia a la actividad o tener ritmos cardíacos anormales, pueden afectar la vida de las personas de muchas maneras. Algunos de estos efectos podrían incluir cómo los adolescentes y adultos jóvenes con cardiopatía de ventrículo único experimentan la escuela, así como sus relaciones con familiares y amigos.
Además, las personas con cardiopatía de ventrículo único necesitan atención médica de por vida y sus experiencias previas de atención médica pueden influir cómo ven sus experiencias de salud actuales y futuras. Para proporcionar la mejor atención posible, es importante que los proveedores de atención médica comprendan cómo los adolescentes y los adultos jóvenes con enfermedad cardíaca de ventrículo único ven sus vidas y experiencias.

¿Qué pasará si mi hijo participa en este estudio de investigación?
Si acepta permitir que su hijo participe en este estudio, le pediremos que:

Antes del estudio:
1. Trabaje con los investigadores para programar una visita en persona para completar el estudio, incluyendo una entrevista y una encuesta escrita.
2. Proporcionar consentimiento informado verbal y escrito para la participación en el estudio el día de la entrevista. La entrevista se llevará a cabo en el campus de UCLA o en su casa si es preferido.

Durante el estudio:

1. Se le pedirá al padre o tutor que firme un formulario de investigación separado de Portabilidad de Seguro de Salud y Responsabilidad (HIPAA) para permitir investigadores revisar los registros médicos de su hijo con respecto a su diagnóstico, tipo y fechas de las cirugías, nivel de saturación de oxígeno, medicamentos y otras condiciones médicas.
2. Completar un formulario de información demográfica y clínica sobre su situación de vida, escuela, estructura familiar y estado médico actual (aproximadamente 20 minutos).
3. Participar en una entrevista con el investigador para hablar sobre sus experiencias en la escuela, relaciones con familiares y amigos, y experiencias de salud. La entrevista será grabada en audio (60 minutos).

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4. Si los investigadores tienen preguntas adicionales que surgan después de la entrevista, pueden contactar a su hijo para programar una entrevista voluntaria adicional, que puede ser hecha en persona o por teléfono.

¿Cuánto tiempo estará mi hijo en el estudio de investigación?
La participación tomará un total de aproximadamente 1 ½ a 2 horas en el día de estudio. Si se necesita una segunda entrevista, ocurrirá dentro de 3 a 5 meses de la primera entrevista.

¿Existe algún riesgo o malestar potencial que mi hijo pueda esperar de esto?
Los posibles riesgos y / o molestias asociados con los procedimientos descritos en este formulario de consentimiento incluyen:

- Puede haber algo de nerviosismo por participar en la entrevista.
- Existe un riesgo muy pequeño de que al hablar durante la entrevista su hijo recuerde los momentos en que se sintieron tristes, asustados o le lastimaron sus sentimientos, y aunque esto es raro, puede hacer que su hijo se sienta incómodo. Tu hijo no tiene que responder a cualquier pregunta que los haga sentir incómodos. Si tu hijo quiere parar durante la entrevista, ya sea por unos minutos o detenerse por completo, pueden hacerlo.
- Existe un riesgo muy pequeño de que se rompa la confidencialidad de su hijo. Se han implementado muchas medidas para evitar que eso suceda. Estas se encuentran en las páginas 3 y 4.

Riesgos y malestares desconocidos:
Los investigadores le informarán a usted y a su hijo si aprenden algo que le pueda hacer cambiar de opinión sobre la participación en el estudio.

¿Hay algún beneficio potencial para mi hijo si él o ella participa?

Su hijo no será beneficiado directamente por su participación en la investigación. Los resultados de la investigación pueden ayudar a los investigadores a aprender más sobre cómo vivir con enfermedad cardíaca congénita compleja y cómo este tipo de enfermedad cardíaca impacta actividades diarias. Esta información puede ayudar en el tratamiento de futuros pacientes con cardiopatía de ventrículo único han tenido un procedimiento de Fontan.

¿Se le pagará a mi hijo por participar?

Su hijo recibirá $50 por participar para compensar costos de viaje y estacionamiento, así como el tiempo y la inconveniencia de participar en el estudio. Se le pedirá a su hijo que firme un recibo del formulario de pago.

¿Se mantendrá confidencial la información sobre la participación de mi hijo?

Los investigadores harán todo lo posible para asegurarse de que la información privada de su hijo sea Mantenida confidencial. La información sobre su hijo será manejada tan confidencialmente como posible, pero participar en la investigación puede implicar una pérdida de privacidad y el potencial de una violación de la confidencialidad. Los datos del estudio estarán asegurados física y electrónicamente. Como con cualquier uso de medios electrónicos para almacenar datos, existe un riesgo de violación de la seguridad de los datos.

Bajo la ley de California, los investigadores no pueden mantener confidencialidad si saben o sospechan razonablemente de incidentes de abuso o negligencia a un niño, adulto dependiente o anciano y debe informar a las autoridades. Esto incluye abuso físico, sexual, emocional, o financiero, o negligencia.

Es posible que los investigadores no puedan mantener confidencialidad si su hijo divulga pensamientos de hacerse daño. Si su hijo revela pensamientos de suicidio o autolesiones usted y su hijo serán remitidos a la sala de emergencias más cercana para recibir una evaluación.

Uso de información personal que pueda identificarle:

Cualquier información que se obtenga en relación con este estudio y que pueda identificar a su hijo permanecerá confidencial. Se divulgará solo con su permiso o según sea necesario por ley. Se mantendrá la confidencialidad asignándole a su hijo un número de identificación único que se utiliza para identificar respuestas escritas y en audio. Los números de identificación del estudio se guardan en un archivo cerrado, separado de los datos del estudio.

Cómo se almacenará la información sobre usted:

Los datos del estudio y los registros se guardan en un archivo cerrado en la oficina del investigador. Los datos electrónicos, incluyendo las entrevistas transcritas, se guardarán en una unidad de computadora protegida con contraseña cifrada.

Personas y agencias que tendrán acceso a su información:

Solo el equipo de investigación tendrá acceso a los datos del estudio.

El equipo de investigación y el personal autorizado de UCLA pueden tener acceso a los datos del estudio y sus registros para monitorizar el estudio. Registros de investigación proporcionados a personal de UCLA no- autorizados no contendrán información identificable sobre su hijo. Publicaciones y / o las presentaciones que resulten de este estudio no identificarán a su hijo por su nombre.

Cuánto tiempo se conservará la información del estudio:

La información del estudio, incluida la lista de códigos de identificadores y cuestionarios, se mantendrá durante tres años después de la finalización del estudio, luego se destruye.

Retiro de la participación por el investigador:

El investigador puede retirar a su hijo de participación en esta investigación si surgen circunstancias que justifican hacerlo. Si es necesario para proteger la salud de su hijo, por ejemplo, si hay angustia visible continua durante la entrevista, es posible que su hijo tenga que abandonar la entrevista, incluso aunque les gustaría continuar. El investigador tomará la decisión y dejará a su hijo saber si no es posible que continúe. La decisión puede ser tomada para proteger la salud y seguridad de su hijo.

¿Cuáles son mis derechos y los de mi hijo si él o ella participa en este estudio?

- Puede elegir si desea o no que su hijo participe en este estudio, y puede retirar su permiso e interrumpir la participación de su hijo en cualquier momento.
- Independientemente de la decisión que tome, no habrá ninguna sanción para usted o su hijo, y no habrá pérdida de beneficios a los que usted o su hijo tenían derecho.
- Su hijo puede negarse a responder cualquier pregunta que no quiera responder y aún permanecen en el estudio. Su hijo puede revisar, editar o borrar la cinta de audio de su entrevista a su solicitud. Si usted o su hijo deciden retirarse del estudio, las cintas de audio y la entrevista transcrita de su hijo serán destruidas. Si su hijo decide editar la cinta de audio de la entrevista, el investigador puede pedirles que lo hagan en otra entrevista grabada.

¿Con quién me puedo comunicar si tengo preguntas sobre este estudio?

- **El equipo de investigación:**
  Si tiene alguna pregunta, comentario o inquietud sobre la investigación, puede hablar con los investigadores. Por favor contacte a la Dra. Nancy Pike al 310-903-2614; npike@sonnet.ucla.edu, o Jennifer Peterson al 714-280-3219; jkpeter1@uci.edu con cualquier pregunta o inquietud sobre la investigación o la participación de su hijo en este estudio.
- **Oficina del Programa de Protección de Investigación Humana (OHRPP) de UCLA:**
  Si tiene preguntas sobre sus derechos como sujeto de investigación o si tiene inquietudes o sugerencias y desea hablar con alguien que no sea los investigadores, usted puede comunicarse con OHLAP de UCLA por teléfono: (310) 206-2040; por correo electrónico: participants@research.ucla.edu o por correo: Box 951406, Los Ángeles, CA 90095-1406.

Contacto para Futuras Investigaciones

Los investigadores pueden contactarme en el futuro para pedirme que participe en otros estudios de investigación de enfermedades del corazón de un solo ventrículo.

☐ SI ☐ NO

Se le entregará una copia de esta información para que la guarde en sus registros.

FIRMA DEL PADRE O TUTOR LEGAL

_____

Nombre del Hijo

_____

Nombre del Padre o Tutor Legal

_____ _____

Firma del Padre o Tutor Legal Fecha

FIRMA DE LA PERSONA OBTENIENDO CONSENTIMIENTO

_____ _____ Nombre de la Persona

Obteniendo Consentimiento Numero de Contacto

_____ _____

Firma de la Persona Obteniendo Consentimiento Fecha

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Appendix G. Minor Assent

UNIVERSITY OF CALIFORNIA, LOS ANGELES ADOLESCENT

(Ages 13-17) ASSENT TO PARTICIPATE IN RESEARCH

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

You are invited to participate in a research study conducted by Dr. Nancy Pike, PhD from the University of California, Los Angeles (UCLA) School of Nursing, and Jennifer Peterson PhD(c) from the School of Nursing at University of California, Irvine. You are invited to participate in this study because you are a teen between the ages of 14 and 17 years with single ventricle heart disease and have had a Fontan operation in the past. Participation in this study is completely voluntary.

Why is this study being done?

The purpose of this study is to better understand the impact of single ventricle heart disease on daily life, including school experiences, relationships with family and friends, and healthcare experiences. Some aspects of having single ventricle heart disease, such as the effects of open heart surgery, being cyanotic (having lower blood oxygen levels), having lower activity tolerance or having abnormal heart rhythms may affect people's lives in many ways. Some of these effects might include how teens and young adults with single ventricle heart disease experience school as well as relationships with family and friends. In addition, people with single ventricle heart disease need life-long medical care and their previous healthcare experiences may influence how they view their current and future healthcare experiences. To provide the best possible care, it is important for healthcare providers to understand how teens and young adults with single ventricle heart disease see their lives and experiences.

What will happen if I take part in this research study?

Please talk this over with your parents before you decide whether or not to participate. We will also ask your parents to give their permission for you to take part in this study. But even if your parents say "yes" you can still decide not to do this.

If you volunteer to participate in this study, the researcher will ask you to do the following:

Before you begin the study:

1. Work with the researchers to schedule an in-person visit to complete the study measures, including an interview and a written survey.
2. Provide verbal and written informed assent (permission) for your study participation on the day of the interview. The interview will take place at the UCLA campus, or your home if preferred.

During the study:

1. Your parent or guardian will be asked to sign a separate Health Insurance Portability and Accountability (HIPAA) research authorization form to allow researchers to review your medical records regarding your heart diagnosis, type and dates of surgeries, oxygen saturation level, medications and other medical conditions.
2. Complete a demographic and clinical information form about your living situation, school, family structure, and current medical status (about 20 minutes).
3. Participate in an interview with the researcher to talk about your experiences in school, relationships with family and friends, and healthcare experiences. The interview will be audio recorded (about 60 minutes).
4. If the researchers have additional questions that arise after the interview, they may contact you to set up an additional voluntary interview, which may be done in-person or by phone.

How long will I be in the research study?

This study will last about 1 ½ to 2 hours on the study day. If a second interview is requested, it will occur within 3 to 5 months of the first interview.

Are there any potential risks or discomforts that I can expect from this study? Known risks and discomforts:

The possible risks and/or discomforts associated with the procedures described in this consent form include:

- There may be some nervousness about participating in the interview.
- There is a very small risk that talking during the interview might make you remember times that you felt sad, scared, or had your feelings hurt, and although this is rare, it may cause you to feel uncomfortable. You do not have to answer any question that makes you uncomfortable. If you want to stop during the interview, either for a few minutes or stop completely, you can do so.
- There is a very small risk that your confidentiality could be broken. There are many measures in place to prevent that from happening, described on page 3 and page 4.

Unknown risks and discomforts:

The researchers will let you know if they learn anything that might make you change your mind about participating in the study.

Are there any potential benefits if I participate?

Possible benefits to me:

You will not directly benefit from your participation in this research.

Possible benefits to others or society:

This study will help the researchers learn more about living with complex congenital heart disease and how this type of heart disease impacts daily activities. This information may help in the treatment of future patients with single ventricle heart disease who have had a Fontan procedure.

Alternatives to participation

If you decide not to take part in this study, or if you withdraw from this study before it is completed, your medical care and relationships with your healthcare providers will not change in any way.

Will I receive any payment if I participate in this study?

You will be given $50 for participation in this study on the study day to compensate you for travel and parking costs as well as your time and inconvenience of participating in the study. You will be asked to sign a receipt of payment form.

Will information about me and my participation be kept confidential?

The researchers will do their best to make sure that your private information is kept confidential. Information about you will be handled as confidentially as possible but participating in research may involve a loss of privacy and the potential for a breach in confidentiality. Study data will be physically and electronically secured. As with any use of electronic means to store data, there is a risk of breach of data security.

Under California law, the researchers cannot keep information about known or reasonably suspected incidents of abuse or neglect of a child, dependent adult, or elder confidential and must report to authorities. This includes physical, sexual, emotional, or financial abuse or neglect.

The researchers may not be able to keep confidential any disclosure of thoughts to harm yourself. If you disclose thoughts of suicide or harming yourself, you will be referred to the closest emergency room for further assessment and evaluation.

Use of personal information that can identify you:
Any information that is obtained in connection with this study and that can identify you will remain confidential. It will be disclosed only with your permission or as required by law. Confidentiality will be maintained by assigning you a unique study identification number that is used to identify your written and audiotaped responses. The key to the study identification numbers is kept in a locked file cabinet, separate from the study data.
How information about you will be stored:
The study data and records are kept in a locked file cabinet in the investigator's office. Electronic data, including transcribed interviews, will be kept on an encrypted, password protected computer drive.
People and agencies that will have access to your information:

Only the research team will have access to the study data.
The research team and authorized UCLA personnel may have access to study data and records to monitor the study. Research records provided to authorized, non-UCLA personnel will not contain identifiable information about you. Publications and/or presentations that result from this study will not identify you by name.

How long information from the study will be kept:
Information from the study, including the code list of identifiers and questionnaires, will be kept for three years after study completion, then destroyed.

- **Withdrawal of participation by the investigator**

The investigator may withdraw you from participating in this research if circumstances arise which warrant doing so. If necessary to protect your health, such as continued visible distress during the interview, you may have to drop out, even if you would like to continue. The investigator will make the decision and let you know if it is not possible for you to continue. The decision may be made to protect your health and safety.

What are my rights if I take part in this study?

You may withdraw your assent at any time and discontinue participation without penalty or loss of benefits to which you were otherwise entitled.

You can choose whether or not you want to be in this study. If you volunteer to be in this study, you may leave the study at any time without consequences of any kind. You are not waiving any of your legal rights if you choose to be in this research study. You may refuse to answer any questions that you do not want to answer and still remain in the study. You may review, edit or erase the audiotape of your interview at your request. If you decide to withdraw from the study, your audiotapes and transcribed interview will be destroyed. If you decide to edit the interview audiotape, the researcher may ask you to do so in another audiotaped interview.

Who can answer questions I might have about this study?

In the event of a research related injury, please immediately contact one of the researchers listed below. If you have any questions, comments or concerns about the research, you can talk to the one of the researchers. Please contact Dr. Nancy Pike at 310-903-2614; npike@sonnet.ucla.edu, or Jennifer Peterson at 714-280-3219; jkpeter1@uci.edu with any questions or concerns about the research or your participation in this study.

If you have questions about your rights as a research subject, or you have concerns or suggestions and you want to talk to someone other than the researchers, you may contact the UCLA OHRPP by phone: (310) 206-2040; by email: participants@research.ucla.edu or by mail: Box 951406, Los Angeles, CA 90095-1406.

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Contact for Future Research

The researchers may contact me in the future to ask me to take part in other single ventricle heart disease research studies.

☐ YES ☐ NO

SIGNATURE OF STUDY PARTICIPANT

I understand the procedures described above. My questions have been answered to my satisfaction, and I agree to participate in this study. I have been given a copy of this form.

_____

Name of Participant

_____ _____

Signature of Participant Date

SIGNATURE OF PERSON OBTAINING ASSENT

In my judgment the participant is voluntarily and knowingly agreeing to participate in this research study.

_____ _____

Name of Person Obtaining Assent Contact Number

_____ _____

Signature of Person Obtaining Assent Date

Protocol ID:IRB#18-001204 UCLA IRB Approved Approval Date: 12/2/2018 Through: 9/19/2021
Committee: South General IRB

Appendix H. HIPAA Release of PHI

IRB# 18-001204

University of California Los Angeles (UCLA) Permission to Use Personal Health Information for Research

Study Title (or IRB Approval Number if study title may breach subject's privacy):
School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

Principal Investigator Name:
Nancy Pike, PhD,

Sponsor/Funding

Agency (if funded):

A. What is the purpose of this form?
State and federal privacy laws protect the use and release of your health information. Under these laws, the University of California or your health care provider cannot release your health information for research purposes unless you give your permission. Your information will be released to the research team which includes the researchers, people hired by the University or the sponsor to do the research and people with authority to oversee the research. If you decide to give your permission and to participate in the study, you must sign this form as well as the Consent Form. This form describes the different ways that the UCLA Health System can share your information with the researcher, research team, sponsor and people with oversight responsibility. The research team will use and protect your information as described in the attached Consent Form. However, once your health information is released by the_____it may not be protected by the privacy laws and might be shared with others. If you have questions, ask a member of the research team.

B. What Personal Health Information will be released?
If you give your permission and sign this form, you are allowing:_____to release the following medical records containing your Personal Health Information. Your Personal Health Information includes health information in your medical records, financial records and other information that can identify you.

Entire Medical Record	Lab & Pathology Reports	Emergency Department Records
Ambulatory Clinic Records	Dental Records	Financial records
Progress Notes	Operative Reports	Imaging Reports
Other Test Reports	Discharge Summary	History & Physical Exams
Other (describe)	Consultations	Psychological Tests

C. Do I have to give my permission for certain specific uses?

Yes. The following information will only be released if you give your specific permission by putting your initials on the line(s).

___I agree to the release of information pertaining to drug and alcohol abuse, diagnosis or treatment.

___I agree to the release of HIV/AIDS testing information.

___I agree to the release of genetic testing information.

___I agree to the release of information pertaining to mental health diagnosis or treatment.

D. Who will disclose and/or receive my Personal Health Information??

Your Personal Health Information may be shared with these people for the following purposes:

1. To the research team for the research described in the attached Consent Form;
2. To others at UC with authority to oversee the research
3. To others who are required by law to review the quality and safety of the research, including: U.S. government agencies, such as the Food and Drug Administration or the Office of Human Research Protections, the research sponsor (insert the name of the sponsor) or the sponsor's representatives including but not limited to (insert the name of the CRO), or government agencies in other countries.

E. How will my Personal Health Information be shared for the research?

If you agree to be in this study, the research team may share your Personal Health Information in the following ways:

1. To perform the research
2. Share it with researchers in the U.S. or other countries;
3. Use it to improve the design of future studies;
4. Share it with business partners of the sponsor; or
5. File applications with U.S. or foreign government agencies to get approval for new drugs or health care products.

F. Am I required to sign this document?

No you are not required to sign this document. You will receive the same clinical care if you do not sign this document. However, if you do not sign the document, you will not be able to participate in this research study.

G. Optional research activity

If the research I am agreeing to participate in has additional optional research activity such as the creation of a database, a tissue repository or other activities, as explained to me in the informed consent process, I understand I can choose to agree to have my information shared for those activities or not.

I agree to allow my information to be disclosed for the additional optional research activities explained in the informed consent process

H. Does my permission expire?

This permission to release your Personal Health Information expires when the research ends and all required study monitoring is over.

I. Can I cancel my permission?

You can cancel your permission at any time. You can do this in two ways. You can write to the researcher or you can ask someone on the research team to give you a form to fill out to cancel your permission. If you cancel your permission, you may no longer be in the research study. You may want to ask someone on the research team if canceling will affect your medical treatment. If you cancel, information that was already collected and disclosed about you may continue to be used for limited purposes. Also, if the law requires it, the sponsor and government agencies may continue to look at your medical records to review the quality or safety of the study.

J. Signature

Subject

If you agree to the use and release of your Personal Health Information, please print your name and sign below. You will be given a signed copy of this form**.**

Subject's Name (print)--*required*

_____

Subject's Signature _____ Date _____

Parent or Legally Authorized Representative

If you agree to the use and release of the above named subject's Personal Health Information, please print your name and sign below.

_____

Parent or Legally Authorized Representative's Name (print) Relationship to the Subject

_____

Parent or Legally Authorized Representative's Signature Date

Witness

If this form is being read to the subject because s/he cannot read the form, a witness must be present and is required to print his/her name and sign here:

Witness' Name (print)

Witness' Signature

Date

Appendix I. HIPAA Release ofPHI-Spanish

Número de Aprobación de la *IRB* 18-001204

Universidad de California Los Angeles (UCLA)
Permiso para utilizar información de salud personal para fines de investigación

Título del estudio (o número de aprobación del IRB si el título del estudio puede violar la privacidad del individuo):

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

Nombre del investigador principal:

Nancy Pike, PhD, RN

Patrocinador/Agencia que aporta la financiación (si está financiado):

A. ¿Cuál es el propósito de este formulario?

Las leyes estatales y federales protegen el uso y la divulgación de su información de salud. Bajo estas leyes, ni la Universidad de California ni su proveedor de atención médica no pueden divulgar su información de salud para fines de investigación a menos que usted dé su permiso. Su información se divulgará al equipo de investigación que está integrado por los investigadores, personas contratadas por la Universidad o por el patrocinador para llevar a cabo el estudio de investigación y personas con autoridad para supervisar el estudio de investigación. Si usted decide dar su permiso y participar en el estudio, tiene que firmar este formulario, además del formulario de consentimiento. Este formulario describe las formas diferentes en que **UCLA Health System** puede compartir su información con el investigador, el equipo de investigación, el patrocinador y las personas responsables de la supervisión. El equipo de investigación utilizará y protegerá su información según se describe en el Formulario de consentimiento adjunto. Sin embargo, una vez que **UCLA Health System** divulgue su información de salud, puede que no esté protegida por las leyes de privacidad y podría compartirse con otros. Si tiene alguna pregunta, hable con un miembro del equipo de investigación.

B. ¿Qué información de salud personal se divulgará?

Si usted da su permiso y firma este formulario, está autorizando a: **UCLA Health System** a divulgar los siguientes expedientes médicos que contienen su Información de salud personal. Su Información de salud personal incluye la información de salud en sus expedientes médicos, registros financieros y otra información que puede identificarle.

☐ Expediente médico	☐ Informes de laboratorio y	☐ Expedientes del departamento de
☐ Expedientes de la clínica	Registros dentales	Registros financieros
Notas de progreso	Informes quirúrgicos	Informes de estudios de imágenes
Otros informes de	Resumen del alta	Historia y exámenes físicos
Otro (describir)	Consultas	Pruebas psicológicas

C. ¿Tengo que dar mi permiso para ciertos usos específicos?

Sí. La información siguiente solo se divulgará si usted da su permiso específico poniendo sus iniciales en las líneas correspondientes.

___Accedo a la divulgación de información relativa al abuso, diagnóstico o tratamiento de drogas y alcohol.

___Accedo a la divulgación de información de pruebas del VIH y del sida.

___Accedo a la divulgación de información de pruebas genéticas.

___Accedo a la divulgación de información relativa al diagnóstico o tratamiento de salud mental.

D. ¿Quién divulgará o recibirá mi Información de salud personal?

Su Información de salud personal podría compartirse con las siguientes personas para los propósitos especificados:

1. El equipo de investigación para realizar el estudio de investigación descrito en el formulario de consentimiento adjunto.
2. Otras personas en UC con autoridad para supervisar el estudio de investigación.
3. Otras personas que estén obligadas por ley a revisar la calidad y seguridad del estudio de investigación, incluyendo: Agencias gubernamentales de los EE.UU., como la Administración de Alimentos y Fármacos (Food and Drug Administration, FDA) o la Oficina para la Protección de Seres Humanos en Estudios de Investigación (Office of Human Research Protections, OHRP), el patrocinador del estudio de investigación (insertar el nombre del patrocinador), o los representantes del patrocinador incluyendo, entre otros, (insertar el nombre del CRO), o agencias gubernamentales de otros países.

E. ¿Cómo se compartirá mi Información de salud personal para realizar el estudio de investigación?

Si accede a participar en este estudio, el equipo de investigación podría compartir o usar su Información de salud personal con las siguientes finalidades:

1. Para realizar el estudio de investigación.
2. Para compartirlo con investigadores en los EE.UU. u otros países.
3. Para mejorar el diseño de estudios en el futuro.
4. Para compartirlo con socios de negocios del patrocinador.
5. Para presentar solicitudes a las agencias del gobierno de los EE.UU. o del extranjero, para obtener la aprobación de nuevos medicamentos o productos de atención médica.

F. ¿Estoy obligado/a a firmar este documento?

No, no está obligado/a a firmar este documento. Recibirá la misma atención clínica si no firma este documento. Sin embargo, si no firma el documento, no podrá participar en este estudio de investigación.

G. Actividad investigadora opcional

Si el estudio de investigación en el que voy a participar por propio consentimiento tiene actividad investigadora opcional adicional, como la creación de una base de datos, un repositorio de tejidos u otras actividades, según se me ha explicado en el proceso de consentimiento informado, entiendo que puedo aceptar que mi información se comparta para dichas actividades o no, a elección mía.

Doy mi autorización para que mi información se divulgue para las actividades investigadoras opcionales adicionales explicadas en el proceso de consentimiento informado.

H. ¿Vence mi permiso?

Este permiso de divulgación de su Información de salud personal vence cuando termine el estudio de investigación y la supervisión requerida del estudio haya concluido.

I. ¿Puedo cancelar mi permiso?

Puede cancelar su permiso en cualquier momento. Puede hacerlo de dos maneras. Puede escribir al investigador o puede pedir a alguien del equipo de investigación que le de un formulario de cancelación de permiso para completarlo. Si cancela su permiso, es posible que no pueda seguir participando en el estudio de investigación. Es buena idea preguntar a alguien del equipo de investigación si la cancelación afectará a su tratamiento médico. Si cancela, la información que ya se había recopilado y divulgado sobre usted podría seguir utilizándose para fines limitados. Además, si la ley así lo exige, el patrocinador y las agencias gubernamentales pueden seguir consultando sus expedientes médicos para revisar la calidad y seguridad del estudio.

J. Firma

Sujeto

Si usted acepta el uso y la divulgación de su Información de salud personal, escriba su nombre en letra de imprenta y firme a continuación. Se le entregará una copia firmada de este formulario.

Nombre del sujeto (en letra de imprenta)--*requerido*

Firma del sujeto	Fecha

Padre/madre o representante legalmente autorizado

Si usted acepta el uso y la divulgación de la Información de salud personal del sujeto antedicho, escriba su nombre en letra de imprenta y firme a continuación.

Nombre del padre/madre o del representante legalmente autorizado (en letra de imprenta)	Relación con el sujeto
Firma del padre/madre o del representante legalmente autorizado	Fecha

Testigo

Si este formulario se le lee al sujeto por no poder este leerlo, debe haber presente un testigo y este debe escribir su nombre en letra de imprenta y firmar aquí:

Nombre del testigo (en letra de imprenta)

Firma del testigo	Fecha

Appendix J. Demographic and Clinical Form

Subject Code #_____ Date: _____

Demographic Information

Please complete the following information to the best of your knowledge. Remember there are no right or wrong answers. If you have any questions or do not understand a question, please ask the investigator or research assistant.

Age: _____

1. Gender:
[ ] Male
[ ] Female
[ ] Non-binary

2. Ethnicity:
[ ] Caucasian / White European
[ ] Latino / Hispanic
[ ] African American
[ ] Asian / Pacific Islander
[ ] Other, please specify: _____

3. Who currently lives with in your home? [check all that apply] [
] Mother [Biologic] only
[ ] Father [Biologic] only
[ ] Both Mother and Father [Biologic] [
] Step-mother or father
[ ] Siblings [brother(s) and /or sister(s)]
[ ] Extended family [grandparents, aunts/uncles, cousins]
[ ] Spouse / Significant Other / Partner
[ ] Children [biologic or adoptive], if so how many_____
[ ] Other _____

4. What is your current marital status?

[ ] Single, living alone
[ ] Single, living with a parent or other family member [
] Single, living with partner
[ ] Married, living alone [separated, widowed, deployment] [
] Married, living with a spouse
[ ] Other _____

5. What is your mother's occupation or job title? _____
[Example: stay at home mom, teacher, sales clerk, cook/ chef, nurse, hairdresser

6. What is your father's occupation or job title? _____
[Example: gardener, electrician, construction worker, cook/chef, doctor, mechanic]

7. What is your current occupation or job title? _____
[Examples: may include student, retired, unemployed, or unable to work due to physical health]

8. What is your highest grade completed?

[ ] < 7th grade
[ ] Junior High School (8th or 9th grade)
[ ] Partial High School (10thor 11th grade)
[ ] High School Graduate
[ ] Partial College (at least 1 year) or specialized training
[ ] College or University Graduate
[ ] Graduate Degree [master's, PhD]

9. What is mother's highest grade level completed?
[ ] < 7th grade
[ ] Junior High School (8th or 9th grade)
[ ] Partial High School (10thor 11th grade)
[ ] High School Graduate
[ ] Partial College (at least 1 year) or specialized training
[ ] College or University Graduate
[ ] Graduate Degree [master's, PhD]

10. What is father's highest grade completed?
[ ] < 7th grade
[ ] Junior High School (8th or 9th grade)
[ ] Partial High School (10thor 11th grade)
[ ] High School Graduate
[ ] Partial College (at least 1 year) or specialized training
[ ] College or University Graduate
[ ] Graduate Degree [master's, PhD]

11. What is your annual household income?
[ ] < $20,000
[ ] $20,000 to less than $30,000
[ ] $30,000 to less than $50,000
[ ] $50,000 to less than $80,000
[ ] $80,000to less than $100,000
[ ] > $100,000
[ ] Unsure
12. What is your zip code?_____.

13. What type of health insurance do you have?

[ ] Private [PPO, HMO]
[ ] Public [Medi-Cal / California Children's Services (CCS), Disability or SSI]
[ ] No insurance / out of pocket or self-pay
[ ] Unsure

13. Do you have any learning, language, reading or attentional issues [if any]? [Please check all that apply]

[ ] None
[ ] Dyslexia [reading disability]
[ ] Language / speech [expressive or receptive, stuttering]
[ ] Attention deficit disorder without hyperactivity [ADD]
[ ] Attention deficit hyperactivity disorder [ADHD]
[ ] Other, specify _____

14. Do you currently, or in the past, had any of these assistances in school? [Please check all that apply to you]

[ ] I have an IEP (Individualized Education Plan) in place [
] I have extra time to complete assignments
[ ] I have extra time to take tests
[ ] I take Special Education classes
[ ] I receive physical, occupational, or speech therapy at school
[ ] I have teaching assistant sit with me during class
[ ] I do not participate in physical education (gym)class

15. What kinds of medical treatments do you currently use? [Please check all that apply to you]

[ ] I take one heart medicine every day
[ ] I take more than one heart medicine every day
[ ] I take at least one other [not heart-related] medicine every day
[ ] I have a pacemaker
[ ] I have an implanted defibrillator [AICD]
[ ] I wear extra oxygen during the day and/or night
[ ] I eat a special diet due to my heart problem
[ ] I have abnormal heart rhythms/palpitations/heart pounding
[ ] I have been told not to do certain physical activities by my doctor [lift weights, run, play contact sports]
[ ] I have been told by my doctor to "self-limit" my activity (stop to rest if I am too tired)

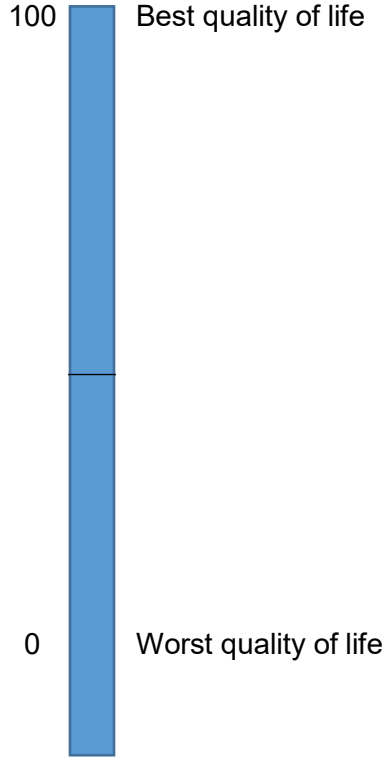
16. In general, would you say that your health is:

[ ] Excellent
[ ] Very good
[ ] Good
[ ] Fair
[ ] Poor

17. Compared to 1 year ago, how would you rate your health in general now?

[ ] Much better now than 1 year ago
[ ] Somewhat better now than 1 year ago
[ ] About the same
[ ] Somewhat worse than 1 year ago
[ ] Much worse than 1 year ago

18. How would you rate your overall quality of life on a scale of 0 to 100 (0 is the worst quality of life and 100 is the best quality of life)?_____(Write the number here)



Appendix K. Chart Review

Chart Review Form for Investigators ONLY

Subject ID #:_____ Date: _____

1. Diagnosis:

   _____

   _____

2. Check Primary Category
   [ ] TA [ ] PA / IVS [ ] HLHS [ ] Ebstein's
   [ ] DILV [ ] DORV
   [ ] Other complex single Ventricle, specify_____

3. Type of Single Ventricle
   [ ] Left [ ] Right [ ] Undetermined

4. Heterotaxy / Asplenia
   [ ] Yes [ ] No

5. Dextrocardia
   [ ] Yes [ ] No

6. Number of Heart Surgeries
   [ ] 1 [ ] 2 [ ] 3 [ ] 4 [ ] ≥ 5

7. Surgery Dates and Names of Operations:
   Date_____ Surgery _____

   Date_____ Surgery _____

   Date_____ Surgery _____

   Date_____ Surgery _____

   Date_____ Surgery _____

8. Catheterization Dates and Name of Procedures [i.e. stents, coils, valves, fenestrations,]:

   Date_____Procedure_____

   Date_____Procedure_____
   Date_____Procedure_____
   Date_____Procedure_____

9. NYHA Class
   [ ] I
   [ ] II
   [ ] III
   [ ] IV

10. LVSF or RVSF%_____(actual number)
    [ ] Qualitative evaluation of function_____[i.e. good, normal, fair, poor]
    [ ] Not available

11. History of Arrhythmias
    [ ] Yes [ ] No
    If yes, type_____

12. Total Number of DAILY Medications [both cardiac and non-cardiac]
    [ ]_____[ ] None

13. List medication names, doses, and how often taken
    _____
    _____
    _____
    _____

14. Cyanotic at Birth
    [ ] Yes [ ] No

15. First surgery before 30 days of life
    [ ] Yes [ ] No

16. Current Oxygen Saturation _____%

17. Current Weight _____kg

18. Current Height _____cm

19. Current BMI _____(calculate)

20. Other medical conditions / surgeries:
    _____
    _____
    _____
    _____

Appendix L. Interview Guide

School and Life Experiences of Adolescents and Young Adults with Single Ventricle Heart Disease

The **specific aims of this study** are to:

1. Explore the perceptions of daily life of adolescents and young adults with SVHD.
2. Examine the influence of SVHD on daily life experiences including school, work, social relationships in adolescents and young adults with SVHD.
3. Explore the influence of SVHD on healthcare experiences of adolescents and young adults with SVHD.

The **research questions** for this study are:

1. What are the lived experiences of school, work, and social relationships of adolescents and young adults with SVHD?
2. What are the successes and challenges associated with school, work and social relationships in adolescents and young adults with SVHD?
3. How do adolescents and young adults with SVHD perceive their quality of life?
4. How do adolescents and young adults with SVHD perceive their health and their healthcare experiences?

Interview Guide

Thank you for agreeing to come in and talk with me about your experiences. As you may know, more and more people with complex heart problems like yours are surviving their congenital heart disease, which makes it very important that healthcare providers better understand what's it's like to grow up with one ventricle, and how congenital heart disease affects your life. It is very impactful to hear directly from the people who are having these experiences. Better understanding might lead to improved ways to help teens and young adults like you deal with and overcome challenges that you face.

There are no right or wrong answers to these questions, because I am trying to understand what your experience is, and how it affects your daily living

I want to remind you about a couple things…Our talk is being recorded with this recorder, but the only identification is your study ID number and the date of the interview. The audio recording will be transcribed into a text file, but that is also only identified with your study ID number and the date. All of these files are completely confidential and only people who have a reason to see them in the study can do so. I won't be offended or upset by anything you tell me, or by any words that you want to use. If you want to take break at any point, that's perfectly OK. Also, if you don't want to answer a particular question, or come back to it later, just tell me and we can do that.

Do you have any questions for me at this point?

Research question 1. What are the lived experiences of school, work, and social relationships of adolescents and young adults with SVHD?

1. Can you please describe a typical day for you? If further prompting needed, How about a typical day at work or school?
    a. How does having SVHD impact your school or work day? How is your day different from your friends or family that don't have heart problems?
    b. How do those daily impacts make you feel?
    c. Has the daily impact of your heart problems changed over time…for example between grade school and high school, or high school and college?
2. How do you get along with your teachers?
    a. Do your teachers know that you have a heart problem?
    b. Do your teachers treat you differently than other students? Can you describe a time when you were treated differently because of your heart problems?
    c. What did that experience mean to you?
    d. How does that impact you and your relationships with teachers?
    e. Some of the other kids we have talked to said that they had some challenges with physical education class or gym class. What is that like for you?
3. How do you get along with your boss and coworkers?
    a. Does your boss or coworkers know that you have heart problems?
    b. Does your boss or coworkers treat you differently than other employees? Can you describe an experience where you were treated differently because of your heart problem?
    c. What did that experience mean to you?
4. Who is the person you can talk to when you are having a bad day, and why is he or she the best person to talk to?
5. Do your friends know about your heart problems?
    a. Do you think that your friends treat you differently because of your heart? Can you give an example of that? How does that make you feel? What does that mean to you?
    b. Do your classmates treat you differently because of your heart? Can you give an example of that? How does that make you feel? What does that mean to you?
    c. How do you tell your friends about your heart problems? What is that like for you? How do they react?
6. You've talked about relationships with friends in school, how about relationships outside of school?
    a. Can you talk about your relationships with brother and sisters?
    b. What about relationships with parents?
    c. How has your heart problem influenced your role in the family? Can you give an example of that? What does that experience mean to you?
    d. Have you been involved in romantic relationships? How has your heart problem influenced that relationship? Can you give an example of that? What did that experience mean to you?

Research question 2. What are the successes and challenges associated with school, work and social relationships in adolescents and young adults with SVHD?

1. Can you describe any limitations that you have in school or work? Physical, educational, activities… Can you describe an example of this?
    a. How have those limitations affected you?
    b. How do those limitations make you feel?
2. What have been challenges that you faced in school or work?
    a. Can you give an example of one of those challenges?
    b. What did that challenge mean to you or make you feel?
    c. How did you meet that challenge?
3. Can you talk about any special classes or help that you need in school or work?
    a. How do those things affect your experiences of school?
4. What would have been helpful to have you succeed in school?
5. What things were not helpful to have you succeed in school?
6. What was your biggest success in school or work? Can you give an example? What made it successful? How did that make you feel? What did that experience mean to you?
7. Can you talk about a time when either you or someone else needed to advocate for you, meaning to speak up or act on your behalf because you needed something or some help that you weren't getting? How does that feel? Do you feel like you can do that for yourself?
8. What challenges do you face in relationships with others because of your heart problems? With friends? With parents? With siblings? With teachers? With co-workers or boss? With romantic partner?
    a. How do you overcome these challenges?

Just to let you know, we're about halfway through the interview, and I really appreciate your willingness to share your experiences. You're helping me to understand what it's like for you, and that's why I wanted to interview you

Research question 3. How do adolescents and young adults with SVHD perceive their quality of life?

1. What things in your life make you happy? Can you give an example? What does this experience mean to you?
2. What would you like your life to be like in the future?
    a. How will you get there?
    b. What challenges and opportunities do you see along the way, and how will you get through them?
3. Compared to people that you know who don't have heart problems that you have, how would you rate your quality of life?
    a. Are there ways that your quality of life is better or worse than others?
    b. Can you give an example of that?

Research question 4. How do adolescents and young adults with SVHD perceive their health and their healthcare experiences?

1. Would you describe yourself as a healthy person? Why or why not?
2. What does the scar from your surgeries mean to you?
3. Compared to others without heart problems like yours, how would you rate your overall health?
    a. How is your health different than others? Can you give an example?
4. Can you describe a really good experience you have had with your health care? What made it a good experience? How did it affect you?
5. Can you describe a bad experience you have had with your health care? What made it a bad experience? How did it affect you?
6. Can you describe your memories of being in the hospital? What did those experiences mean to you? How did those experiences affect you?
7. If you could give doctors and nurses advice about taking care of and interacting with people who have the same kinds of heart problems as you have, what would that advice be? What about teachers?
8. If you have made the transition from kids heart clinic to adult heart clinic, what was that like for you? If you haven't yet made that transition, how do you think that will go?

From your experiences, what else should I know about growing up with heart problems?

We've reached the end of the interview. Thank you so much for sharing your experiences with me, I think that what we have talked about will be very helpful to improve understanding of what school and other life experiences are like for people with single ventricle heart disease. Do you have any questions for me? While we are analyzing data in the next few months, it's possible that we may come up with some other questions or need clarification on something that we talked about. If that happens may I contact you? Would you prefer a phone call or an email? The questions or clarification would probably be something we could do by phone if that is more convenient for you.