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Antecedents and Outcomes of Self-Care in Adults with Congenital Heart Disease

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An abstract of
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Abstract

Antecedents and Outcomes of Self-Care in Adults with Congenital Heart Disease

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Background: Adults with congenital heart defects (ACHD) face long-term complications related to prior surgery, abnormal anatomy, and acquired cardiovascular conditions. Although self-care is an important part of chronic illness management, few studies have explored ACHD self-care.

Purpose: The purpose of this study was to describe the antecedents and outcomes of self-care in the ACHD population, guided by the Family and Self-Care Framework.

Methods: Persons with moderate or severe ACHD were recruited from a single ACHD center. Self-care (maintenance, monitoring and management behaviors), antecedents (individual, clinical, knowledge, behavioral, and family-related factors), and outcomes (functional capacity, symptom burden, and 3-month health resource use) were collected via self-report, chart review, and a six-minute walk test (6MWT). Bivariate correlations, linear and logistic regression were used for analyses.

Results: A total of 132 persons were included (mean age 36 ± 11 years, 55% female, 81% Caucasian, 54% severe ACHD). Only 44.7%, 27.3%, and 23.3% of participants performed adequate self-care maintenance, monitoring and management, respectively. In regression analysis, higher self-efficacy, higher education, male gender, higher perceived family support, and more comorbidities explained 25% of the variance in self-care maintenance (R^2=.248, F(5, 123)=9.44, p<.001). Older age, higher depressive symptoms, higher self-efficacy, and higher NYHA Class explained 23% of the variance in self-care monitoring (R^2=.232, F(2, 124)=10.66, p<.001). Higher self-efficacy and higher NYHA Class explained 9% of the variance in self-care management (R^2=.094, F(2, 80)=5.27, p=.007). Participants walked an average of 1365 ± 275 feet on the 6MWT and experienced 7.4 ± 4.8 symptoms. A total of 101 (80.8%) participants experienced 310 healthcare encounters (275 office visits, 21 hospitalizations, 20 ED visits). Self-care management (β=-44.20, p=.001) and monitoring (β=-14.59, p=.03) were independently associated with functional capacity; self-care maintenance and monitoring were independently associated with symptom burden (β=-.39, p<.001; β=1.52, p<.001); higher self-care monitoring was an independent predictor of ≥ 1 health encounter (OR=1.21, 95% CI [1.09, 1.35], p<.001).

Conclusions: Suboptimal self-care is common among persons with ACHD, and is associated with worse functional health. Modifiable factors of self-efficacy and perceived family support may be important targets of future self-care interventions to improve health outcomes in the ACHD population.

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Table of Contents

CHAPTER ONE

Introduction

Statement of the Problem

Fifty years ago only one third of children born with congenital heart disease survived to 10 years of age, but today 90% of children survive to adulthood and over one million adults are living with congenital heart disease (ACHD).[1,2] Although the majority of adults had surgical repair in childhood, they are not cured. Surgical sequelae and residual defects cause late complications such as arrhythmia, heart failure, and the development of renal, pulmonary, and hepatic comorbidities.[3,4] With the exception of those with mild heart defects, complications are universal and require lifelong surveillance and illness management. Although self-care is a vital component of chronic illness management, little is understood about the practice of self-care in ACHD. Limited evidence of specific behaviors indicates that low levels of self-care may be a significant problem for persons with ACHD. Between 40-60% of adults, for instance, are not engaged in routine care and up to 85% do not meet physical activity recommendations.[5] Further, at least 20% of women do not attend pre-pregnancy counseling despite high rates of maternal and fetal complications.[6-8] Evidence also suggests that low levels of self-care are associated with poor health outcomes. In ACHD, those with a three-year lapse in care are over three times more likely to require emergent interventions, receive additional diagnoses, and experience more symptoms,[9] and those who are less physically active have greater reductions in functional capacity.[10] Thus, improving self-care may be an effective means of reducing ACHD morbidity, but the development of interventions to promote ACHD self-care is limited by the lack of evidence regarding antecedent factors of self-care in this population.

In the similar acquired heart failure (HF) population, various individual, clinical, and behavioral antecedent factors are known to influence self-care,[11] but few factors have been identified in ACHD. The studies that do exist disagree on the individual and clinical factors they identify to explain self-care activities in this population.[12-14] Furthermore, no studies of self-care in an adult cohort of patients with congenital heart disease have examined the role of knowledge and behavioral characteristics such as depression.[15,16] In addition, family influences are increasingly recognized as important determinants of self-care across populations,[17] and may contribute uniquely in survivors of childhood illness through parent-child relationships.[18]

Purpose

The purpose of this study is to determine the antecedents of self-care in ACHD in the family context, and to explore the relationship between self-care and health outcomes that have been previously identified in this population (functional capacity),[19] or unexplored but identified as important in the similar acquired HF population (symptom burden and health resource use (HRU)).[20-22] By identifying antecedent factors related to ACHD self-care and potential health outcomes to target for improvement, this study is a first-step towards developing interventions to promote self-care and improve health outcomes in the ACHD population.

Specific Aims

Specific Aim 1: Determine the individual (age, race, gender, education level, insurance status, marital status), clinical (heart defect severity, NYHA classification, number of comorbidities), ACHD knowledge, behavioral (depressive symptoms and self-care self-efficacy), and family (parental overprotection and self-care support) antecedent factors of self-care in ACHD.

RQ1: What is the relationship between individual and clinical antecedent factors and level of self-care (as measured by the Self-Care Index for ACHD)?

H1: Lower levels of self-care will be associated with knowledge, behavioral, and family antecedent factors of:

a. Lower levels of ACHD knowledge (as measured by the ACHD Health Passport).
b. Higher levels of depressive symptoms (as measured by the Patient Health Questionnaire-9).
c. Lower levels of self-care self-efficacy (as measured by the Self-Care Confidence Scale).
d. Lower levels of perceived support from family (as measured by the Stanford Self-Efficacy Scale).

H2: Parental overprotection (as measured by the Parental Bonding Instrument) will moderate the relationship between behavioral antecedent factors and level of self-care.

Specific Aim 2: Explore the relationship between self-care and health outcomes (functional capacity, symptom burden, and HRU) in ACHD.

H2: Controlling for heart defect severity and number of comorbidities, lower levels of self-care will be associated with:

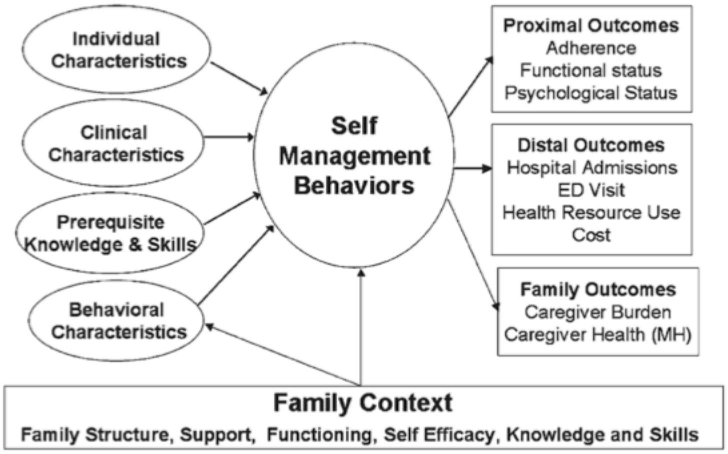
a. Lower functional capacity (as measured by the Six Minute Walk Test).
b. Higher symptom burden (as measured by the Congenital Heart Disease Symptom Survey).
c. Greater health resource use (defined as provider office visits, ED visits, hospitalizations, within 90 days of recruitment).

Conceptual Framework

Self-care is one of the oldest and most central concepts in the field of nursing. The concept of self-care was first introduced in the 1970s by Dorothea Orem in her Theory of Self-Care Deficit as an individual's ability to care for self versus need for care provided by a nurse in times of illness.[23] The concept has evolved over time with the increasing burden and complexity of chronic illness care in the United States. Greater emphasis is placed on the role the individual with the chronic illness and support systems have expanded from the nurse only to family, a variety of healthcare providers, and the structure of the health system. Though no one definition of self-care is agreed upon, self-care generally involves the ability of persons to care for themselves, that is, perform actions and make decisions required to maintain health and manage limitations or complications due to illness.[24]

Many theoretical frameworks have been developed to explain the reasons persons with chronic illness perform high or low levels of self-care, what factors are most important to successful self-care, and what health outcomes are expected to improve through self-care. Frameworks may be either generic or disease-specific and have been based on a wide variety of health behavior theories such as socioecological models, health belief model, theory of planned behavior, and health promotion theory.[25] In the ACHD population, no theoretical frameworks currently exist to guide self-care research. Given its holistic perspective and utility in the similar acquired HF population, this study will test Dunbar *et al's* (2008) Family and Self-Care Framework (FSCF) for use in the ACHD population (Figure 1).[17]

Figure 1. Family and Self-Care Framework



The FSCF was originally developed from a synthesis of literature on self-care in chronic illnesses and HF, the role of family in chronic illness management, and self determination theory (SDT).[17] In this framework, self-care is defined as "activities that involve self-care decisions and actions around the recommended HF self-care behaviors" (p.5).[17] Primary concepts include individual characteristics, clinical characteristics, prerequisite knowledge and skills, and behavioral characteristics, self-care behaviors, health outcomes (proximal, distal, family), and the family context. Individual characteristics include sociodemographic factors and health beliefs; clinical characteristics include disease severity, cognitive status, comorbidities, and self-care complexity; knowledge and skills refer to prerequisite understanding of self-care behaviors and associated skills; behavioral characteristics include self-efficacy, mood states, and motivation. Health outcomes resulting from self-care include proximal outcomes (treatment adherence, functional health status, and psychological health status), distal outcomes (HRU and costs), and family outcomes (caregiver burden, caregiver health). The family context includes family structure, family knowledge and skills, family functioning, and family support.

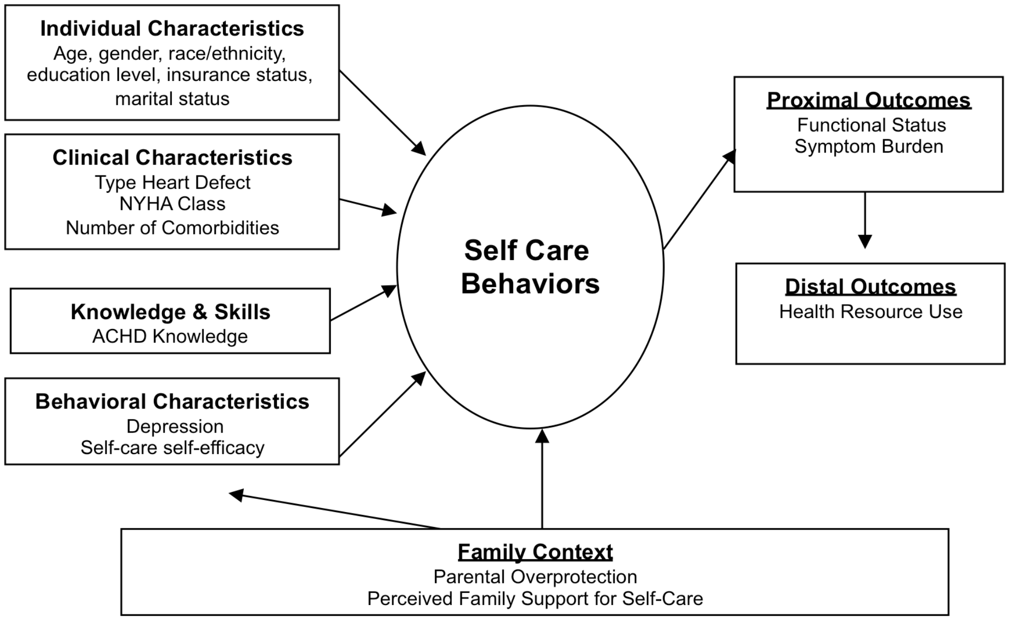
The primary propositions in this framework include the following: 1) Level of self-care behavior is dependent upon multiple domains including individual characteristics, clinical characteristics, prerequisite knowledge and skills, and behavioral characteristics; 2) The family context may directly influence self-care behaviors or indirectly influence them through behavioral characteristics; 3) Level of self-care behavior has a direct influence on health outcomes, including level of adherence, functional status, psychological status, HRU and costs, and family outcomes (caregiver burden and caregiver health). The primary assumptions of this framework are that self-care directly influences health outcomes, and that family is essential to self-care. Family not only influences level of self-care, but is also an integral part of the process of self-care. Thus, health outcomes are reserved not only for the individual, but also the family. This framework also assumes that motivation for self-care is driven by influences of SDT. This theory of human motivation assumes persons are motivated by both intrinsic and extrinsic sources, and the sources that promote autonomy, competence, and relatedness have the greatest influence on motivation.[26] The primary manner in which this framework incorporates SDT is the ability of autonomy supportive behavior by family to increase individual motivation for self-care.[27]

Several advantages exist for using the FSCF to guide the aims and design of this study. First, empirical evidence supports the inclusion of family in the self-care process.[17,28] In acquired HF, support from family has been associated with better medication adherence and lower readmission rates, [29,30] and interventions to improve family support of self-care have been shown to improve adherence to sodium restrictions.[27] Family may also be particularly important for adults born with a life-long chronic illness. In the adult cystic fibrosis population, qualitative studies report the importance of continued support from parents for self-care as well as the potential negative influence parents and family may have on self-care through controlling behavior.[31] This finding is not surprising and may be similar for persons with ACHD. For most adults, family plays the primary role in self-care for almost two decades, and maybe longer. The transition to primary responsibility for care may be thus delayed or impeded. The other primary advantage of the FSCF is its identification of specific, measurable antecedents of self-care that have been previously associated with self-care in the HF population.[11]

Although acquired HF and ACHD are similar disease processes, they are not identical, and the ACHD population is generally a younger population. Thus, there is a possibility that the FSCF does not include all factors that influence self-care in the ACHD population. Another limitation of the FSCF is its exclusion of factors outside the individual and family that may influence self-care such as the healthcare system and the environment. Problems with the healthcare system, such as lack of reimbursement for self-care education and support, communication and trust of healthcare providers, and lack of coordination between care providers in persons with comorbid conditions, may impede an individual's ability to carry out self-care.[11,32] Future studies should include these factors to determine their salience in the ACHD population. Lastly, the FSCF provides specific variables to explain level of self-care, but it lacks some direction in which variables are the most salient and the potential interconnectedness between variables such as the known relationship between disease severity and depression.[33]

In the ACHD population, little attention has yet been given to examining specific antecedent factors that may promote or hinder self-care and no framework to guide research has been developed. Although the FSCF has some limitations, its holistic perspective, utility in the acquired HF population, and inclusion of family factors makes it a strong framework for guiding the aims of this study. Specific individual, clinical, knowledge/skills, behavioral, and family antecedent characteristics of self-care and related health outcomes used in this study are outlined in Figure 2.

Figure 2. Family and Self-Care Framework for ACHD



Relevance of Study

Congenital heart disease is the most common birth defect occurring in 8:1,000 live births.[34] Advances in medicine and surgical techniques over the past four decades have dramatically reduced childhood mortality and resulted in an adult population now exceeding one million and growing by 5% each year.[2,35] Associated healthcare use is also rising quickly, with a doubling of hospitalizations and quadrupling of healthcare costs between 1998 and 2005.[36] Approximately 500,000 adults with moderate to severe ACHD represent the highest burden of morbidity and are the focus of this study.[37] Common diagnoses include Tetralogy of Fallot, Transposition of the Great Arteries, and single-ventricle physiology palliated with a Fontan procedure. Though most adults with moderate to severe ACHD had surgical repair in childhood, they must manage varying but significant complications due to unique anatomy and consequences of childhood surgery.[38] The most common complications include HF, arrhythmia, and the need for reoperation.[3] Hepatic, pulmonary, and renal comorbidities, as well as risk for protein-losing enteropathy and thromboembolic events may also occur.[3,39,40] Many are at risk for sudden death, and 75% of adults with heart defects die of defect-related causes.[41,42] Both the personal and societal burden of ACHD is rising. A significant need exists to understand ways to prevent adverse health outcomes in this population. The practice of self-care in ACHD is an understudied but potentially critical means of reducing adverse health outcomes.

BACKGROUND

Self-Care Defined

Self-care is defined in accordance with the FSCF as “activities that involve self-care decisions and actions around the recommended [HF] self-care behaviors” (p.5).[16] These self-care activities are operationally defined according to Riegel *et al* as 1) self-care maintenance (treatment adherence activities), 2) self-care monitoring (symptom monitoring activities) and 3) self-care management (activities related to symptom response).[43] Guidelines for self-care in ACHD do not currently exist. However, several guidelines for the medical management of ACHD have been published including the American College of Cardiology/American Heart Association (AHA) guidelines for the management of ACHD, the AHA Guidelines for the management of transition to ACHD, and annual Betheseda conference guidelines.[38,44,45] Based on these published guidelines and expert ACHD clinician practices, important self-care behaviors were identified.

Self-Care Maintenance

Similar to the acquired HF population, diet, weight management, alcohol and tobacco restriction, exercise, immunizations, and advance directives are considered important self-care maintenance behaviors in persons with ACHD. Additional self-care behaviors specific to ACHD include preventive behaviors related to endocarditis, pregnancy, and complication of anesthesia.[38] Adults with cyanotic defects or mechanical devices are at increased risk for endocarditis and require antibiotics prior to invasive procedures including dental work. Dental hygiene, avoiding body piercings and tattoos are also preventive behaviors.[46] All women with ACHD are at risk for pregnancy complications, though risk increases by severity of ACHD.[47] Thus, birth control and knowing pregnancy risks are important parts of self-care in this population. Many adults, especially those with single ventricle physiology are at high anesthesia risk due to an inability to augment blood flow and require advanced planning to prevent adverse outcomes.[48] The ability to provide a description of heart defect and surgeries with non-ACHD providers has also been identified as an important maintenance behavior, as many non-ACHD providers lack of familiarity with ACHD.[38] Medication adherence is recommended, but evidence-based treatment in ACHD is rare, and the relative contribution of adherence is generally unknown.[38]

Self-Care Management

The most commonly studied self-care behavior in this population is routine follow-up, as lapses in care are widespread.[12,13,49,50] Follow-up is typically considered a maintenance behavior, and a large portion of persons with ACHD may have asymptomatic disease progression or a latent period between surgical correction and late complications. Further, for adults with life-long symptoms or limitations, recognition of and response to worsening symptoms is more difficult. Managing disease, thus, includes regular cardiac screening. Management issues and recommendations also vary by type of heart defect or late complication. For instance, adults with arrhythmias must learn to manage palpitations, malfunctions in pacemakers, and exercise limitations.

Self-Care in ACHD

Evidence suggests that low levels of certain self-care behaviors are widespread in ACHD. Between 40-60% of adults are considered lost to follow up care,[8,9] up to 85% do not meet physical activity,[5] and at least 20% of women do not attend pre-pregnancy counseling despite high rates of pregnancy complications.[6-8] In a study of adults' understanding of endocarditis, less than 50% had adequate understanding of their risk and prevention behaviors.[51] Among young adults with ACHD, one study has shown that over 50% reported smoking or binge drinking at least once, though other studies representing a wider age range of adults have reported rates of smoking and drinking alcohol relatively low compared to the general population.[52-54] Other self-care behaviors such as medication adherence, symptom monitoring, and immunization uptake have surprisingly no known documentation in ACHD.

Although all late complications cannot be prevented, there is evidence that inadequate levels of self-care are associated with an increase in ACHD morbidity. Compared to adults with ACHD who engage in routine care, those who have a three-year lapse in care are over 3 times more likely to require emergent interventions, over 12 times more likely to receive a new diagnosis, and almost 5 times more likely to be symptomatic.[9] Also, adults with ACHD who are less physically active have greater reductions in functional capacity.[10] These outcomes of increased HRU and symptoms, and reduced functional capacity are also associated with self-care in the acquired HF population in which interventions to improve self-care have been shown to improve these outcomes. Thus, it is anticipated that these outcomes will be important targets of improvement in ACHD self-care as well.

Antecedents of Self-Care

Low levels of certain self-care behaviors have been described in the literature, yet specific antecedent factors that may promote or hinder self-care in the ACHD population have not been examined. Given its holistic perspective and utility in the acquired HF population, this study will test the Dunbar *et al's* Family and Self-Care in HF Framework for use in the ACHD population. The framework, as described previously, is a synthesis from the literature on self-care in chronic illnesses and HF, and self determination theory.[26,17] The framework includes individual, clinical, knowledge, skills, and behavioral antecedent characteristics of self-care, self-care behaviors, and related health outcomes in a family context. Characteristics and outcomes considered in this study are described in Figure 2 and reviewed as follows.

Individual Characteristics

Potential demographic factors related to self-care include age, gender, race or ethnicity, educational level, insurance status, and marital status. In an unpublished dissertation of adolescents and young adults with heart defects, older age but not gender or race was associated with higher levels self-care.[14] In studies of adults not engaged in routine care, demographic antecedents vary with no consensus on their relative importance in predicting behavior.[12,13] Persons with higher levels of education and who are in supportive marriages are known to have higher levels of self-care in acquired HF,[55-57]but their influence in ACHD self-care is unknown. Lack of insurance is known to influence self-care through access to care,[58] and lack of insurance has been cited as a primary reason persons with ACHD have lapses in routine care.[59]

Clinical Characteristics

Potential <u>clinical characteristics</u> related to self-care include ACHD disease severity and comorbidities. ACHD severity has been associated with physical activity levels in ACHD, but relationship to overall self-care is unknown.[5] Comorbidities increase the burden of self-care through complex medication regimens, difficulty distinguishing symptoms, and disease management conflicts.[11] In the acquired HF population, comorbidities are associated with lower medication and diet adherence, difficulty interpreting healthcare professionals instructions for multiple illnesses, and decreased self-care confidence.[11,60,61] In addition to risk for illness such as diabetes and hypertension due to age, adults experience other comorbidities related to pulmonary, renal, and hepatic function.[62] Comorbidities are expected to influence self-care, but the relationship is untested in ACHD.

Knowledge

Although knowledge may be insufficient to explain level of self-care, it is recognized as an important prerequisite.[11] In a study of young adults with ACHD, the majority knew something about their heart condition, but only 36% had a clear understanding of their heart defect.[63] Another study found that young adults with ACHD were characterized by lack of knowledge and awareness about future health risks due to their heart defect.[64] Surprisingly, one ACHD center found that 35% of adults could not name their defect, and 50% did not understand the reason for endocarditis and pregnancy related risks.[65] In the development of a health knowledge questionnaire for ACHD, Moons *et al* found that patients generally understood their ACHD, however they had poor understanding of the reasons for follow up and the symptoms of decreased heart function.[51] Another study by Van Deyk *et al* confirms these findings, with less than half of patients understanding the need for regular follow up to detect clinical deterioration.[66] In a qualitative analysis of self-care in adolescents and young adults, Fleck et al found that those who could explain their heart defect in layman's terms had higher levels of self-care.[14] However, the relationship between knowledge and self-care in ACHD remains poorly understood.

Behavioral Characteristics

Depression

In ACHD, between 22-54% of persons have either self-reported or clinical depression.[16,15] Notably, in a sample of persons with ACHD considered to be well adjusted, 36% were found to have diagnosable depression.[67] Rates of depression do vary by country, but North American populations report higher rates than European populations.[68,69] The negative relationship between self-care and depression has been well-established in persons with acquired HF,[11] but the relationship between self-care and depression in ACHD is unknown. Given the high rates of depression in ACHD, it is anticipated that depression may be an important factor negatively influencing self-care behaviors.

Self-Care Self-Efficacy

Self-efficacy is a central construct in Bandura's social cognitive theory (1977) and is defined as the "conviction that one can successfully execute [a given] behavior required to produce [a given] outcome (p. 193).[70] Though self-efficacy has been operationalized as a global construct indicating general confidence is one's ability to cope with any given situation, this operationalization is not supported by Bandura's theory, in which self-efficacy is expected to vary by behavior within the same individual.[71,72] According to Bandura's theory persons with higher levels of perceived self-efficacy for a given behavior are more likely to engage in the behavior. The use of self-efficacy in self-care research has been supported by Bandura.[73] Self-care self-efficacy is defined as the perceived ability to successfully engage in self-care behaviors and decisions.[74] Self-care self-efficacy has been shown to be predictive of self-care behavior across multiple chronic illnesses, including the similar populations of cystic fibrosis (CF) and acquired HF.[74,75,76] In acquired HF, self-efficacy has been shown to influence self-care behaviors, and interventions improving self-efficacy have demonstrated improvements in self-care.[75,76] Indeed, self-care self-efficacy has consistently been shown to be one of the strongest predictor of HF self-care behaviors.[77,78]

Recently, self-care self-efficacy was significantly related to level of self-care in young adults with ACHD even after adjusting for multiple clinical and psychosocial variables.[14] In regards to the particular behavior of exercise in ACHD, two studies have shown that low levels of exercise were associated with lower exercise self-efficacy.[5,79] It is anticipated that self-care self-efficacy will be an important predictor of self-care in the ACHD population, but this remains unconfirmed.

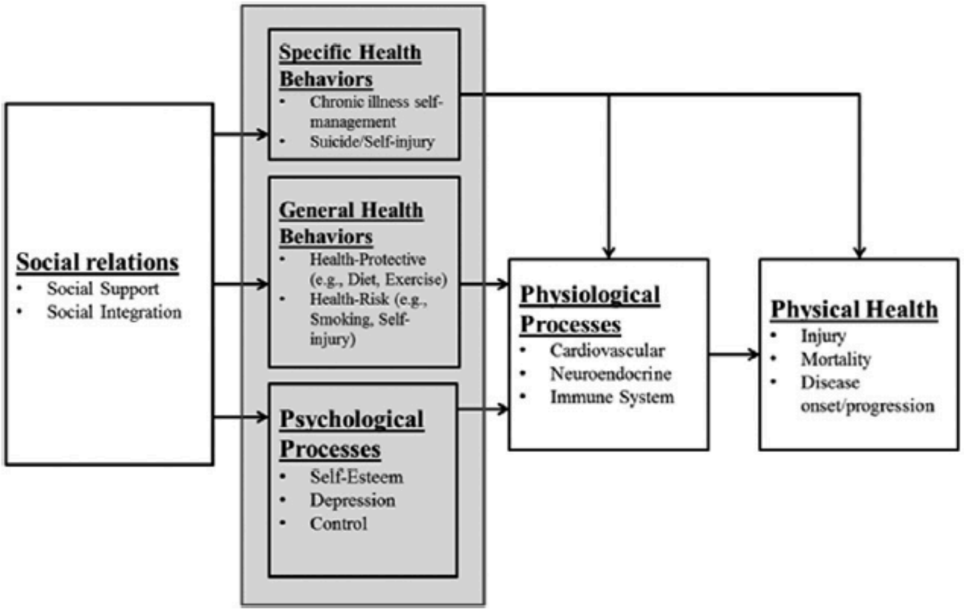
Family Context and Social Support

Beyond the fact that social support plays an important role in self-care[80,81] and family plays a central role,[17,28] the evidence is surprisingly inconsistent about the types of social support that are most influential in persons with chronic illness.[82] In the Social Relations and Health Model based on the classic stress-buffering hypothesis of Cohen & Willis (1985) and more recent work incorporating physiological processes, Tay *et al* (2013) describe the relationship between social relations and health (Figure 2).[83] Social relations include both social support and social integration. Social support is defined as either received or perceived support and is commonly categorized into instrumental or tangible, informational, and emotional support.[84] Social integration refers to the types, frequency, and number of social contacts without regard for their perceived benefit. The distinction between these two concepts, in addition to measurement variability, has been cited as a reason for a lack of consistency regarding the role of social support in health outcomes.[85] For example, in persons with acquired HF, being married (social integration) is not as influential as the quality of the marriage relationship (social support) on self-care.[17] This is likely due to the instrumental support that spouses offer such as assistance with symptom management, treatment adherence, and decision-making.[56,57] To assume that a non-significant finding between marital status and self-care translates into a non-significant relationship between social support and self-care would be misguided. Although social integration is known to predict disease progression and mortality, social support--and in particularly instrumental support--has been reported as more predictive of self-care than social integration across multiple chronic conditions.[83]

According the Social Relations and Health Model (Figure 3), social relations may have a direct or indirect influence on self-care. Indirectly, social support or social integration may influence self-care through an interaction with psychological processes, including mood states and self-efficacy. In the ACHD population, little is understood about how social relations influence self-care. One qualitative analysis reported that social support from both family and friends were important for effective self-care.[14] Due to this finding, the role of family support across chronic illnesses including other populations of adult survivors of childhood illness,[31] and in working with the Family and Self-Care Framework,[17] social support for self-care from family will be measured in this study.

The role of the family is increasingly recognized as an important factor influencing level of self-care in chronic illness,[17] and in an unpublished dissertation, family functioning in ACHD was associated with level of self-care.[14] In acquired HF, support from family has been associated with better medication

Figure 3. Social Relations and Health[83]



adherence and lower readmission rates,[29,30] and interventions to improve family support of self-care have been shown to improve adherence to dietary sodium restrictions.[27] Though support from family is generally positive, it has also been shown to have a negative influence on self-care through critical or controlling behavior.[28,81,86] According to Durkheim's classic theoretical definition of social integration, the structure of social relationships such as parenthood provide a sense of meaning in life as well as a set of controls on individual behavior.[84] Parental overprotection refers to when these controls may become unhealthy, marked by '"intrusion, excessive contact, infantilization, and prevention of independent behavior"' (p.261).[18] Children growing up with a chronic illness are known to experience more controlling parental behaviors than healthy subjects.[87] Parental overprotection has lasting effects into adulthood including poorer social, psychological, and behavioral functioning.[88,89] In persons with ACHD, parental overprotection in childhood has been associated with symptoms of anxiety, health distress, and activity limitations.[18,90] The concept of parental overprotection aligns well with the Family and Self-Care Framework because it is part of the family context and hinders autonomy supportive behaviors.[17]

Apart from specific health risk behaviors, the direct relationship between parental overprotection and level of self-care has not been explored in adult populations. Based on evidence regarding its relationship to psychosocial outcomes in ACHD and self-care in adolescent populations, it is hypothesized that parental overprotection may have a direct influence on self-care or it may moderate the relationship between behavioral characteristics (depressive symptoms, self-efficacy) and self-care. The hypothesized influence on self-care may be bidirectional. Though parental overprotection may lead to reduced independence, lower autonomy support, and thus lower levels of self-care, it may also be associated with higher levels of self-care. This phenomenon is well documented in adolescents with chronic illnesses when parental overprotection leads to increased treatment adherence due to increased oversight of treatments.[91,92] Further, in adults with cystic fibrosis, it has been reported that persons who believe their illness is controlled by family or a healthcare provider have higher levels of self-care than those who believe they are in control of their illness because pleasing parents or providers serves as a motivating factor.[31] Parents of persons with ACHD are reported to frequently attend clinic appointments with their adult children.[93] This may indicate that persons with ACHD have higher levels of self-care due to continued parental involvement.

However, the degree and directionality of parental overprotection in persons with ACHD remains unknown.

Proximal and Distal Outcomes

Functional Capacity

Functional capacity is operationally defined as exercise tolerance. Exercise tolerance is almost universally decreased in moderate to severe ACHD and clinical symptoms usually include exertional dyspnea and fatigue.[94] There is evidence that daily physical activity is moderately associated with exercise capacity (r=.40), and some evidence that increasing self-care behaviors such as physical activity can increase functional capacity in ACHD.[19,95] Certain medications such as ace inhibitors and pulmonary vasodilators have demonstrated increased functional capacity in ACHD, but data are limited and the relative contribution of medication adherence is unknown.[94] Functional capacity is among the few known predictors of poor health outcomes in ACHD, especially those with single ventricle physiologies who represent a small but severely affected portion of the population.[96,97]

Symptom Burden

Symptoms are recognized to be an individual's subjective experience of changes in their physical or mental state.[98] Symptom burden is less clearly defined in the literature, but often referred to and defined in this study, as the average perceived frequency, severity, and distress of symptoms.[99,100] Although knowledge of the symptom burden in ACHD is limited, the evidence suggests that many experience a considerable number of symptoms. In a small sample of adults with severe ACHD, most frequently experienced symptoms were found to be shortness of breath with ambulation, dizziness, and palpitations.[101] In a larger ACHD sample, frequent symptoms included shortness of breath with activity, excessive perspiration, palpitations, dizziness, and nocturnal polyuria, with females experiencing symptoms more frequently and with greater distress.[102] In the acquired HF population, symptom burden increases with poor self-care such as a lack of adherence to sodium or fluid restrictions,[21,99] but the relationship between symptom burden and self-care behaviors has not been explored in ACHD and symptom burden is understudied.

Health Resource Use

HRU is increasing rapidly in the ACHD population. Between 1998 and 2005, the number of ACHD hospitalizations in the U.S. doubled, growing from 36,0000 to 72,000 admissions, and the total estimated healthcare spending increased from $691 million to $3.16 billion.[36] The most recent data from 2007 show an additional 10% increase in hospitalizations.[103] Little information is available regarding emergency room usage in this population, although it is known that an increase in usage occurs in young adulthood.[104] HRU is expected to continue to increase with survival of adults with the most severe types of heart defects, and the 5% expected growth in the ACHD population each year.[35] Limited but negative associations between self-care and HRU have been reported. Poor routine follow-up, for instance, has been associated with the need for emergent interventions.[9] In the similar acquired HF population, interventions to improve self-care have been shown to reduce hospitalizations by up to 66%.[105]

In ACHD, the increase in HRU does parallel the increase in number of adults, but it remains unknown if self-care behaviors may be an important, modifiable predictor.

Discussion

In grand scheme of adult heart disease, persons with ACHD do not represent a majority of costs or numbers, but they are a growing population and a particularly vulnerable one. The infrastructure of the current healthcare system is not well equipped to care for them. Over half a million adults with moderate to severe CHD require specialty care and about 100 specialty centers exist in the United States. These numbers indicate that only about 10% of adults who require specialty care are receiving it.[106,107] One impediment to the lack of specialty centers is the lack of ACHD-specialty providers. General adult cardiologist are not equipped to care for persons with ACHD, but only 500-600 ACHD-specialty providers exist in the United States.[107] A new board certification for ACHD subspecialty was just approved in 2012 by the American Medical Association with hopes to improve access to quality care for persons with ACHD. Lack of access is certainly a contributing explanation for the 40-60% of persons with ACHD are considered lost to follow-up care.[8,9] Participating in routine care is particularly important in this population as disease progression often occurs asymptomatically, and studies have shown that persons with gaps in care may have poorer outcomes.[9] Further, persons with ACHD may live a far distance from an ACHD specialty center and must rely on less well-trained primary care doctors or cardiologists, increasing these adults' need to understand their own disease management. In light of the vulnerability of poor access to care, understanding how to support self-care is even more critical.

A recent review of policies in ACHD reflected on the fact that CHD is the single most successfully treated form of heart disease. Survival rates to adulthood changed from less than 50% to 90% in just four decades.[108] The federal government has invested significant dollars into pediatric CHD that has led to this success, but it has also led to a growing, vulnerable population of adults with CHD. Federal investment is needed to remember these children and continue their CHD success story into adulthood by determining means of reducing morbidity and mortality. Self-care, as described previously, represents an understudied but potentially important means to reduce morbidity in this population. Federal investment is needed to better understand the self-care abilities, needs, and support structure for person with ACHD. This dissertation study represents a first step and is among the first federally funded projects focused on ACHD self-care. This study will help reduce the gap in knowledge regarding the practice of self-care in ACHD, particularly in relation to the various antecedent factors that may be important for self-care and the health outcomes that may be influenced by self-care including symptom burden, functional capacity, and HRU.

Methods

Research Design

A cross-sectional, descriptive design was used to examine the relationship between antecedent factors (individual, clinical, behavior, knowledge, and family factors) and self-care, and self-care and proximal health outcomes (functional capacity and symptom burden) in persons with moderate to severe ACHD, and a descriptive, prospective component was used to examine the relationship between self-care and the distal health outcome of HRU. Data collection was conducted at four time points including at enrollment and monthly for three months.

Sample

Inclusion criteria were adults who are ≥18 years, have a moderate or severe ACHD diagnosis, NYHA class II-III, and are English-speaking. *Exclusion criteria* were 1) developmental delay impairing ability to independently complete surveys, 2) adult diagnosis of ACHD, and potential confounders of a walking test including 3) current pregnancy, 4) disability preventing ability to walk, and 5) less than one month post-op from cardiac surgery. The sample size to achieve adequate statistical power to address all aims of this study was based on the most complex aim, specific aim one. G*Power software was used to determine sample size and based on plans for linear regression models to detect a medium effect size for the change in R^2 of the predictors.[109] This effect size was determined based on previous studies using the Self-Care in Heart Failure Index and similar antecedent factors, with R^2s ranging from 0.15-0.40.[77,110,111] A sample size of 111 achieves 80% power at an alpha of 0.05 to detect a change in R^2 of 0.20 for 16 predictors per the projected statistical procedures and was the target sample size. Other projected sample sizes based on an alpha of 0.05 are included in Table 1.

Table 1. Sample Size Projections

	R^2=0.15	R^2=0.20	R^2=0.30
85% Power	157	121	86
80% Power	143	111	79

Setting

The Nell Hodgson Woodruff School of Nursing served as the coordinating center for this study. Participants were recruited from the Emory Adult Congenital Heart Center, one of the largest ACHD specialty clinics in the United States.[107] In the past two years, approximately 2,000 unique patient visits have occurred (53% female, 66% Caucasian, and 67% between 20-39 years old), with the majority of patients seen having moderate to severe ACHD.

Measurement

The following variables and measures were used:

Self-care Index for ACHD (SCI-ACHD). The SCI-ACHD is a revised version of the Self-Care in HF Index.[43,74] This 32-item measure of self-care was revised by the investigator with the original author's permission by adding items related to ACHD self care including endocarditis prevention, birth control, and ACHD symptoms, and deleting some items unrelated to ACHD self-care such as salt and fluid restrictions.[38] The SCI-ACHD includes independently scored subscales of self-care maintenance, self-care monitoring, self-care management, and self-care confidence in accordance with self-care of chronic illness theory.[112] A 4-point Likert scale is used to assess items, from 1=(Never/Not Likely/Not Confident) to 4=(Always/Very Likely/Extremely Confident). SCI-ACHD face validity was assessed by 4 ACHD experts (2 expert patients and 2 ACHD providers) for relevance and completeness; 3 additional ACHD providers completed formal content validity testing in accordance with guidelines outlined by Polit et al.[113] Items without complete agreement (n=9 for clarity, n=2 for relevance) were either removed or changed to satisfy critiques. In pilot testing, as expected with the small sample size, Cronbach's alpha (CA) was low for all subscales (0.41 maintenance, 0.76 monitoring, and 0.52 management), but scores had good variability and patients indicated that the instrument was readable and clear. To further validate scores in this study, medical chart review were used to determine concurrent validity with routine ACHD follow-up behavior and flu vaccination in the year prior to enrollment in the study. Adequate follow-up was defined as at least one appointment in the year prior to enrollment in accordance with the AHA follow-up guidelines for ACHD.[38] Flu vaccination is routinely documented in compliance with organizational standards.

Demographics and Clinical Variables: A self-reported questionnaire was used to collect age, gender, race, educational level, marital status, and insurance status. Comorbidities (number and type) and type of heart defect were collected via medical chart review. Type of heart defect was classified as moderate or severe based on AHA guidelines.[38]

ACHD Knowledge Survey is a 22-item, investigator-developed survey based on the ACHD Patient Health Passport, a 7-page booklet used to store personal health information, including health history, health risks, and self-care needs.[114] All questions are scored as correct or incorrect, summed for total score and then standardized on a 100-point scale. Higher scores reflect higher levels of knowledge.

Patient Health Questionnaire-9 (PHQ-9) is a widely used and well-validated 9-item, self-reported scale measuring depression.[115] Items are scored on a 4-point Likert scale (0=not at all to 4=nearly every day) with a total score ranging from 0 to 27. Scores 5-9 indicated mild depression, 10-14 indicate moderate depression, 15-19 indicate moderate-severe depression, and scores >19 indicate severe depression symptoms. Internal consistency reliability is excellent in ACHD populations (CA=0.88).[116]

Self-Care Self-Efficacy

The 6-item self-care confidence subscale of the SCI-ACHD was used to determine perceived self-care skills. In the HF population internal consistency reliability is excellent (CA=0.86).[43] Pilot testing of the instrument in 30 ACHD patients showed anticipated low reliability (CA=0.47) due to small sample size, but good variability and acceptability in patients.

Six Minute Walk Test (6WMT) is a widely used objective measure of functional capacity in acquired HF and has moderate to strong correlations with cardiopulmonary exercise testing (CPET).[117,118] The relationship between CPET and the 6MWT has not been well studied in ACHD, however the test has been used in ACHD patients with pulmonary arterial hypertension comorbidity or post-Fontan palliation to measure exercise capacity, and is responsive to medication trials.[119-121] Given its low cost, highly feasible clinical administration, and association with daily activity abilities, the 6MWT was used in this study.[122] The test was given in accordance with American Thoracic Society guidelines.[122] Participants are asked to walk as far as possible along a level, marked hallway for six minutes. Heart rate, blood pressure, respiratory rate, and oxygen saturations are monitored immediately prior to and after the six-minute walk test. Participants must have a resting heart rate of less than 120 beats per minute at the time to participate. Participants with cyanotic heart defects, defined as resting oxygen saturations less than 85%, had continuous oxygen saturation monitoring. Baseline saturation less than 65% or a drop in saturations greater than 15% from baseline stopped the test in accordance with clinical safety guidelines. The test was stopped immediately for symptoms of chest pain, intolerable dyspnea, leg cramps, ashen appearance, diaphoresis, or staggering. Total distance walked was used in analysis.

Congenital Heart Disease Symptom Survey (CHDSS) is a 24-item revision of the Heart Failure Symptom Survey by the investigator with the original author's permission by adding or removing symptoms as relevant to ACHD.[100] The scale asks persons to rate the frequency and severity of symptoms and the symptom's affect on enjoyment of life over a one-month timeframe, on a 5-point Likert scale. The average score of frequency, severity, and affect on enjoyment of life are used to create a symptom burden score for each symptom, and summed for an overall symptom burden. CHDSS face validity was assessed by 4 ACHD experts (2 expert patients and 2 ACHD providers) for relevance and completeness; 3 additional ACHD providers completed formal content validity testing in accordance with guidelines outlined by Polit et al.[113] Items without complete agreement were either removed or changed to satisfy critiques (n=8 clarity, n=9 relevance). Preliminary pilot testing in 30 ACHD patients showed excellent internal consistency reliabilities with a CA of 0.92 (frequency), 0.93 (severity), and 0.92 (enjoyment). Further psychometrics were tested in this study.

HRU Questionnaire is a self-reported diary of planned or unplanned health care services consumed. Physician visits, ED visits, and hospitalizations during the study were recorded. Self-report was essential as many patients live far from the specialty center and receive care at facilities other than Emory Healthcare. Health care services consumed and hospital length of stay at Emory Healthcare was verified through electronic medical records. Total use for each variable was obtained by summing reported episodes. Reason for the contact was categorized.

Family Self-care Self-efficacy: Perceived support for self-care from family was measured using the Stanford Chronic Illness Family, Friends, and Community Self-Efficacy Scale. This 4-item questionnaire measures perceived support for self-care and has adequate reliability (CA=.77, ICC=.85) across multiple chronic illnesses, but has not yet been validated in the ACHD population.[123] Language related to friends and community was removed from questions so that family remained the only support mechanism, consistent with the variable to be measured.

Parental Overprotection was measured using the Parenting Bonding Inventory Overprotectiveness subscale,[124] a 13-item scale with excellent internal consistency reliability in the ACHD population (Cronbach's alpha=0.89).[18] Adults are asked to rate the attitudes and behaviors of parents in the first 16 years of life. Each item is measured on a 4-point Likert scale from 0=very unlikely to 3=very likely and a total score is summed. Higher score reflect higher overprotectiveness.

Table 2. Self-reported instruments

Construct	Instrument	N. Items	Max. Expected Time (min)
Self-Care	SCI-ACHD	32	10
Demographics	Standard	10	5
Knowledge	ACHD Knowledge Survey	10	10
Depression	PHQ-9	9	5
Self-care Self-efficacy	SCI-ACHD	-	-
Symptom Burden	CHDSS	24	10
Family self-care support	Stanford Self-Efficacy Scale	4	3
Parental Overprotection	Parental Bonding Instrument	23	10
TOTAL		112	53

Data Collection and Procedures

After obtaining a partial HIPAA waiver, potential participants were identified through medical chart review. All patients meeting eligibility requirements and having an upcoming office visit were mailed a letter of invitation. Screening for recruitment occurred monthly on an ongoing basis until the total sample size was achieved. The letter of invitation included a welcome letter, consent, HIPAA form, and packet of questionnaires. In the welcome letter, patients were advised of the study and offered an opportunity to be screened via telephone for inclusion. They had the opportunity to decline further involvement at that time by not contacting the PI or not following up on the information provided. The PI only followed-up by telephone one time, a week from the date the letter was sent. Patients could decline at anytime and had the chance to be removed from the mailing list thru their request. Patients initiating contact as a result of the mailed invitation were screened by telephone by the PI to confirm eligibility, further explain the study, and answer any questions. A verbal consent was taken over the telephone, and signed, written consent and a HIPAA form was obtained at the clinic visit. Patients filled out questionnaires on paper or electronically through a link to a secured REDCap database (see Appendix A), prior to the clinic visit. Patients could revoke consent at any time.

Eligible patients could also be approached directly at a clinic visit by physician referral. In this case, after identifying herself, the PI asked permission to describe the study to the patient. If the subject agreed, a one-minute script about the study was presented after which those interested in participating were given a chance to read the informed consent and ask questions. Written informed consent was obtained prior to data collection. Patients completed surveys in a designated quiet space such as a conference room, which were then reviewed by the PI for completeness. For all patients, the six-minute walk test was completed immediately following the clinic visit. Total time to complete all testing was approximately 45-60 minutes. Participants were given a HRU diary to fill out prospectively. HRU data was collected via telephone by the PI or research assistant or submitted electronically by the patient through a secured REDCap database at 30, 60, and 90 days. At time of recruitment, participants were compensated with a free valet parking token and $10 gift card. On completion of HRU diary, participants were mailed a $10 gift card.

Protection of Human Subjects

Risk to Human Subjects

Participation in the proposed study was voluntary and participants could refuse any part of the data collection or withdraw at any time without consequences to themselves, families, or communities. The involvement of human subjects in this study included research participants' completion of surveys related to their perceived physical and mental health, knowledge of their heart condition, symptoms, and family relationships, a six-minute walk test of functional capacity, and tracking of HRU for 3 months, as well as provide permission for access to their medical records.

The study was designed to determine the antecedent factors and health outcomes related to self-care in adults with congenital heart disease (ACHD). The study was limited to adults with moderate to severe heart defects as defined by the AHA due to their high burden of morbidity compared to those with mild heart defects, and to adults experiencing NYHA class II-III symptoms, as symptom burden is a primary outcome in this study and some adults are asymptomatic. Age is limited to adults, ages 18-85 years. A target sample size of 108 was chosen based on the fewest number of participants needed to achieve adequate statistical power to answer specific aims 1 and 2.

Participants were recruited from Emory's Adult Congenital Heart (EACH) Center. With Emory IRB approval and a partial HIPAA waiver, potential participants were identified through screening electronic medical records for patients with upcoming office visits. Potential participants were approached, with permission of the attending healthcare provider, at clinic visits and given an outline of the study criteria and procedures. Informed consent was signed and surveys and six-minute walk test was conducted immediately after clinic visit. In order to meet sample size requirements for the aims of this study, a convenience sample was obtained, with every qualifying person approached for inclusion in the study. Every effort was made to recruit based on targeted enrollment demographics (see Table 3).

Sources of Materials

The research material that was obtained from all participants included the following: demographics, clinical data, surveys, and a six-minute walk test of functional capacity. Demographics included age, gender, race, marital status, education level, and insurance status. Clinical data included medical chart review for type of CHD, NYHA class, comorbidities, and last annual flu shot. Surveys included self-care in ACHD, ACHD knowledge, depressive symptoms, and self-care confidence. Six-minute walk test was conducted as outlined in the research proposal in accordance with American Thoracic Society guidelines.[91]

Finally, the electronic medical record was reviewed for a period of 3 months following enrollment to verify reported HRU from the patient.

Standardized protocols were established to collect and manage data in order to protect participant confidentiality. All data were collected electronically via Research Electronic Data Capture (REDCap), a secure web-based data collection tool. Participants filled out surveys directly into REDCap on an electronic tablet device. All participants were given a unique identification number and only the PI and sponsor had access to identifiable information given REDCap's ability to have varying level of access to data. Thus, shared data with biostatistician or any consultant only included de-indentified data. REDCap is a password-protected database maintained by Emory University and backed up instantaneously. All electronic data will be de-identified after ten years in accordance with Emory University policy. Publications resulting from this study will not name or describe individual participants in an identifiable way.

Potential Risks

Minimal risk to subjects was anticipated including the time inconvenience of filling out surveys (30 minutes) and physical distress (fatigue, heart-related symptoms) from completing the six-minute walk test (15 minutes). Breaks could be taken at any time at the participant's request. Psychological risk included distress from increasing awareness of ACHD or depressive symptoms. Physical stress or fatigue could be experienced through participating in the six-minute walk test, but American Thoracic Society guidelines were followed explicitly. Participants did not complete the test if they did not meet vital sign and symptom criteria. Further, a registered nurse performed all testing and observe participants closely for fatigue or other symptoms. No financial or legal risks were associated with participation in this study.

Recruitment and Informed Consent

Potential participants were identified through electronic medical chart review. Potential participants were approached at an office visit at the EACH Center with permission of the attending healthcare provider. Informed consent was obtained from all participants prior to beginning the study and data collection occurred immediately following the office visit.

Protection Against Risks

This study was designed to minimize and protect against potential risks. Informed consent was obtained prior to collecting any data on participants and participants were ensured that participation or no participation in the study would not affect the care receive at the EACH Center. The PI is appropriately trained and certified in human subjects research, and in conduction of six-minute walk tests. As previously described, participant confidentiality was protected by electronic collection and storage of all data in REDCap. Only the PI and sponsor have access to identifiable data. All other consultants only have access to de-identified data. Raw data will not be shared without a data sharing agreement.

Adverse events were not anticipated, but any events were reviewed and reported promptly to the Emory IRB. In the event that a participant experienced emotional distress, they were referred to an ACHD healthcare provider or clinical psychologist available to ACHD patients. Further, the total PHQ-9 was reviewed during data collection. Though the PHQ-9 is not able to make a diagnosis of depression, it is a useful screening instrument. **If the PHQ-9 had a response ≥ 10**, indicating moderate or greater depression, they were informed and if acceptable, their results shared with their ACHD healthcare provider. They were referred to their provider for follow-up. **If the PHQ-9 form had a response of ≥ 20 OR at least 5 items with selections in the 2 right columns, one of which is either #1 or #2,** indicating moderate to severe depression, the participant was referred to the Georgia behavioral health link (1-800-715-4225), a 24-hour crisis and access line for psychiatric referrals. Resources included brief clinical screening and triage, setting up psychiatric appointments for participants, and mobile crisis teams. **If the PHQ-9 form had a response ≥ 16 and has expressed suicidal ideations,** the ACHD healthcare provider was informed immediately and referral for emergency psychiatric-mental health assistance was made. This protocol for depression screening was explained prior to consent.

To prevent physical distress during the six-minute walk test, the American Thoracic Society guidelines were followed explicitly. The PI has extensive experience conducting six-minute walk tests in both the ACHD and acquired heart failure populations. Six-minute walk tests were conducted at the clinic, with ACHD healthcare providers present to assess a participant experiencing physical distress and with the medical equipment and supplies needed to treat a participant, though this was not anticipated due to the self-limiting nature of the test.

Potential Benefit of Proposed Research to Subjects and Others

Direct potential benefits were not anticipated in this study, but benefit to others with ACHD was anticipated as a result of this research study. The results were used to design and test interventions to improve self-care in ACHD, which was anticipated to improve health outcomes in this population. Participants received a small compensation for the time taken to participate in this study.

Importance of the Knowledge to be Gained

The proposed study will help reduce the gap in knowledge regarding the practice of self-care in ACHD, particularly in relation to the various antecedent factors that may be important for self-care and the health outcomes that may be influenced by self-care including symptom burden, functional capacity, and HRU. Little is understood about the antecedents and outcomes related to self-care in ACHD, and the knowledge gained from this study will support clinical guidelines and interventions regarding self-care in ACHD.

Inclusion of Women and Minorities

Targeted enrollment of women and minorities is based on a combination of EACH Center demographics (53% female, 66% white, 16.4% black, 1.1% Hispanic or Latino, 0.4% Asian, 11.1% unrecorded, and 5.4% other/declined) and the congenital heart defects birth rate demographics in the metropolitan Atlanta area (50.2% female, 52.1% white, 36.0% black, and 11.9% other).[94] Thus, the targeted enrollment for this study includes 50% female, 66% white, 26% black, 8% other, and 3% Hispanic or Latino. Gender, race, and ethnicity were monitored and recruitment strategies would be altered to ensure adequate representation of women and minorities.

Inclusion of Children

Since the NIH definition of children includes anyone less than age 21 years, the proposed study will include those ages 18-20 years. In Georgia, legal age for consent to participate in research is 18 years. At the EACH Center 3.3% of the population is less than 20 years, so every effort was made to proportionally represent these ages in the proposed study (n=4).

Table 3. Targeted/Planned Enrollment

Targeted/Planned Enrollment Table

TARGETED/PLANNED ENROLLMENT: Number of Subjects			
Ethnic Category	**Sex/Gender**		
	Females	**Males**	**Total**
Hispanic or Latino	2	2	4
Not Hispanic or Latino	52	52	104
Ethnic Category: Total of All Subjects *	54	54	108
Racial Categories			
American Indian/Alaska Native	1	0	1
Asian	2	2	4
Native Hawaiian or Other Pacific Islander	0	1	1
Black or African American	17	17	34
White	34	34	68
Racial Categories: Total of All Subjects *	54	54	108

Statistical Analysis

All instruments were scored in accordance with author guidelines. Descriptive statistics were analyzed for all study variables with examination of type and extent of missing data, although little missing data was expected due the primary cross-sectional nature of the research design and data collection procedures. However, any missing data were checked for any bias (predictors of missing) and assessed for assumptions of MCAR (missing completely at random) and MAR (missing at random). Data were reviewed for normality assumptions and outliers, as well as multicollinearity using tolerance and variance inflation factors and condition index. In the presence of significant multicollinearity, stepwise variable selection methods were used to select the optimal set of variables within each sequential block of the regression models. All data were analyzed using SPSS version 21 and an alpha set at 0.05.

<u>Specific Aim 1</u>: Determine the <u>individual</u> (age, race, gender, education level, insurance status, marital status), <u>clinical</u> (heart defect severity, NYHA classification, number of comorbidities), <u>ACHD knowledge</u>, <u>behavioral</u> (depressive symptoms and self-care self-efficacy), and <u>family</u> (parental overprotection and perceived family support) antecedent factors of self-care in ACHD. Bivariate relationships between individual, clinical, knowledge, behavioral, and family antecedent factors and level of self-care were determined using Pearson’s correlation. Multiple stepwise regression analysis was used to statistically evaluate the relationship between antecedent factors and level of self-care.

The moderating effect of parental overprotection on level of self-care will also be statistically evaluated. Moderators are variables that change the strength and/or direction of the relationship between two variables and are tested for statistically through interaction terms.[125] In this instance, the interaction term between parental overprotection and self-care self-efficacy or depression was added to the regression model to see if it achieved significance. If the term is statistically significant, a moderation affect is likely, and the direction and size of the coefficient may be evaluated for strength of moderation. The use of parental overprotection as a moderator is based on the known relationship with psychological outcomes in the ACHD population.[18]

<u>Specific Aim 2</u>: Explore the relationship between self-care and health outcomes (functional capacity, symptom burden, and HRU) in ACHD. Bivariate relationships between level of self-care and health outcomes (functional status, symptom burden, and HRU) was determined using Pearson's correlation. Multiple stepwise regression analysis was used to statistically evaluate the relationship between level of self-care and each health outcome, controlling for severity of ACHD, and number of comorbidities.

Limitations

Although this study will fill an important gap in understanding of self-care in ACHD, it is not without limitations. First, validated measures of self-care and symptom burden in ACHD have not been previously established in the literature. Although the self-care and symptom burden instruments developed by the researcher in this study are based on well-validated instruments in the acquired HF population and show preliminary reliability and validity in pilot testing, it is possible that they may require additional revisions to improve reliability and validity. Further psychometric testing of both instruments was conducted in this study. Second, a cross-sectional and descriptive design does not allow for the determination of cause and effect relationships, but it does allow for the exploration of the relationships among many potential antecedent factors of self-care to generate future hypotheses and guide intervention development. Also, the prospective collection of HRU information will allow for cause and effect relationships to be explored, but a 90-day time frame may not be long enough to capture adequate HRU for analysis. Finally, participants were recruited from one ACHD center, potentially limiting generalizability of findings and causing selection bias as some adults may be followed in non-specialty centers. Every effort was made to recruit a diverse sample that reflects population demographics of the specialty center.

Summary

The epidemiology of congenital heart disease has changed dramatically over the last four decades, with the burden of shifting from the acute, life-threatening illness of the newborn to the chronic, life-limiting illness of the adult.[2] Evidence-based treatment and management of ACHD remains in its early stages, with the first AHA/ACC guidelines published just four years ago.[38] The role of self-care, a vital component of chronic illness management, also remains evidence-scarce in this population. This study is an important first step in expanding the evidence-base for the antecedent factors and outcomes associated with self-care in ACHD that will lead to the development of evidence-based interventions supporting self-care in ACHD and reducing both the personal and societal burden of ACHD. Three manuscripts written for publication in a peer-reviewed journal are included in this dissertation including a detailed, updated literature synthesis on ACHD self-care (Chapter Two), and two manuscripts addressing the specific aims of this study (Chapter Three and Four). A comprehensive, integrative review of the results of this dissertations study, as well as implications for future research, practice, and policy are included in Chapter Five.

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CHAPTER 2: Self-Care in Adults with Congenital Heart Disease: A Literature Synthesis

Abstract

Background: Adults with congenital heart disease (ACHD) are an emerging population of chronically ill adults who face significant morbidities due to residual defects and surgical sequelae. Self-care is an essential component of chronic illness management, but relatively little is known about the practice of ACHD self-care or its contribution to health outcomes. Objective: The purpose of this literature synthesis is to describe and identify gaps in what is currently known about ACHD self-care including 1) the adequacy of self-care behaviors, 3) antecedents of self-care, and 4) health outcomes related to self-care. Methods: A review of the literature guided by the Family and Self-Care Framework and ACHD practice guidelines was conducted from July 2013-February 2015. Results: No published guidelines or comprehensive conceptual framework currently exists on the topic of ACHD self-care. Thirty-five studies of specific self-care behaviors including physical activity, routine follow-up care, substance use (alcohol, tobacco, illicit drugs), pregnancy-related behavior, weight management, and endocarditis prevention were identified, but no studies were identified on overall self-care. Significant gaps in knowledge exist, including the description of key self-care behaviors such as medication adherence and symptom monitoring, as well as the antecedents and outcomes of ACHD self-care including family influences. Conclusions: Self-care is an understudied but potentially important mechanism to improve health outcomes in the ACHD

population. Further research is warranted to establish ACHD self-care guidelines and identify the health outcomes for which self-care is a modifiable antecedent.

Introduction

Congenital heart disease is the most common birth defect occurring in 8:1,000 live births or approximately 40,000 children every year.[1] Advances in medicine and surgical techniques over the past four decades have dramatically reduced childhood mortality and resulted in an adult population now exceeding one million.[2,3] With the exception of persons with mild defects, congenital heart disease is a chronic condition that requires lifelong surveillance and illness management. The majority of adults had surgical repair in childhood, but they are not cured. Surgical sequelae and residual defects cause late complications such as heart failure, arrhythmia, and the development of multi-organ comorbidities, which often require reintervention or reoperation.[4,5] Further, as adults with congenital heart disease (ACHD) age, they have additive contributions from acquired cardiovascular conditions such as hypertension and coronary artery disease.[6] Many adults are also at risk for sudden death and 75% of persons with ACHD die of cardiac-related causes.[7,8] In parallel, associated HRU is increasing rapidly. Between 1998 and 2005, the number of hospitalizations doubled and healthcare costs quadrupled to $3.16 billion.[9] The number of ACHD hospitalizations increased an additional 10% in 2007,[10] and is only expected to escalate with the 5% annual growth of the ACHD population and the now normative survival of persons with severe types of ACHD.[3]

Although self-care is considered an essential component of chronic illness management, very little is understood about the practice of self-care in the ACHD population. Indeed, no known studies have been published on the topic of ACHD self-care and unlike the acquired heart failure (HF) population,[11] no self-care guidelines have been developed. Limited evidence of specific self-care behaviors, however, suggests that inadequate ACHD self-care may be a significant problem and associated with poorer outcomes. An estimated 40-60% of persons with ACHD for instance, are not engaged in routine cardiac follow-up;[12,13] a three-year lapse in cardiac care has been associated with three times the likelihood of requiring emergent interventions, receiving additional diagnoses, and experiencing more symptoms.[13] Lower levels of physical activity have been associated with reduced exercise capacity and quality of life, [14] yet between 39-100% of persons with ACHD do not meet physical activity recommendations.[15-21] Further, at least 20% of women do not attend pre-pregnancy counseling despite higher rates of maternal and fetal complications.[12,22,23] Thus, improving self-care may be an effective means of reducing morbidity in the growing ACHD population. The purpose of this review is to describe what is currently known about ACHD self-care including 1) the adequacy of self-care behaviors, 3) antecedents of self-care, and 4) health outcomes related to self-care. Synthesizing current knowledge of ACHD self-care will allow a solid foundation for establishing guidelines, research priorities, and clinical standards that can be targeted through interventions to improve self-care and consequently, health outcomes, in the ACHD population.

Conceptual Framework

Since no guidelines or theoretical frameworks currently exist for ACHD self-care, Dunbar *et al's* Family and Self-Care (FSC) Framework was chosen *a* *priori* to guide this review of the literature (Figure 1).[24] The FSC Framework was originally developed in 2008 by Dunbar *et al* from a synthesis of literature on self-care in chronic illnesses and HF, the role of family in chronic illness management, and self determination theory.[24] In this framework, self-care is defined as “activities that involve self-care decisions and actions around the recommended (HF) self-care behaviors” (p.5).[24] Self-care activities are defined operationally according to Riegel *et al* as 1) self-care maintenance (treatment adherence or any other activities that maintain physiologic stability), 2) self-care monitoring (symptom monitoring activities) and 3) self-care management (symptom response activities).[25] The FSC Framework views self-care as a complex phenomenon with its success influenced by multiple antecedent domains including individual and clinical characteristics, prerequisite knowledge and skills, and behavioral characteristics such as self-efficacy, depressive symptoms, and health beliefs. Importantly, the FSC Framework recognizes the importance of accounting for the family context of self-care, which may directly influence self-care behaviors or indirectly influence them through behavioral characteristics. The family context includes family structure, family functioning, and family support. Health outcomes resulting from self-care include proximal outcomes (symptom burden, functional and psychological health status), distal outcomes (mortality, quality of life, HRU and costs), and family outcomes (caregiver burden, caregiver health). Although multiple frameworks for self-care in chronic illness exist, the FSC Framework was chosen for its holistic perspective, utility in the similar acquired HF population, and its emphasis on the role of family in self-care that may be particularly important for adults living with a chronic condition since birth.[26]

Methods

An extensive review of the literature was conducted to examine and synthesize current literature on ACHD self-care. Articles were identified by the following databases: PubMED, CINAHL, and Web of Science. The search was conducted from December 2014 to February 2015. No article-type restrictions were used. Age range was limited to adults only (19+), publication date to the past ten years and language to English. Primary search terms included major mesh term "*congenital heart defects"* with *self-care, self-management,* and *chronic illness management.* This search produced no published articles on the topic of overall ACHD self-care. To further explore current evidence on ACHD self-care, specific self-care behaviors were identified from published ACHD management guidelines including the American College of Cardiology/American Heart Association (ACC/AHA) guidelines for the management of ACHD, and the AHA Guidelines for the management of transition to ACHD, the 32nd Betheseda conference guidelines, and clinical experts.[27-29] Search terms related to these self-care behaviors and the process of selection are summarized in Figure 2. In addition, bibliographies of selected articles were searched. The level of scientific evidence (strong, moderate, or inconsistent) for antecedents and outcomes of specific self-care behaviors was determined using a best evidence synthesis based on level of evidence described from Cochrane Collaboration review groups (Table 1).[30]

ACHD Self-Care Behaviors

A total of 35 published articles were identified on ACHD self-care behaviors including physical activity, routine follow-up care, substance use (alcohol, tobacco, illicit drugs), endocarditis prevention, weight management, and pregnancy-related behavior (Table 2), and discussed by frequency of studies. The majority of studies focused on physical activity and routine follow-up. ***Physical activity*** is considered an important ACHD self-care behavior, as functional capacity is almost universally decreased in persons with ACHD.[31] Physical activity also confers well established benefits to general cardiovascular health, skeletal muscle function, immune function, obesity prevention, and psychological or cognitive function.[32] For the majority of persons with ACHD, physical activity recommendations are the same as the general public, that is, performing muscle strengthening activities at least 2 times per week and 75 minutes of vigorous or 150 minutes of moderate activity per week, with each activity session lasting at least 10 minutes.[32] Special populations of ACHD included persons with cyanosis, arrhythmia, or ventricular dysfunction require individualized recommendations,[32] but all persons, even persons with comorbid pulmonary arterial hypertension, can safely engage in some level of physical activity.[33] Studies indicate that anywhere from 39-100% of persons with ACHD have either low levels of physical activity or are not active at all.[15-21] A small to moderate, positive relationship (r=.23-.44) between daily physical activity level and functional capacity, typically measured in terms of peak oxygen uptake, has been reported in several studies.[15,18-21] Further, although limited to pilot studies, increasing physical activity levels can improve functional capacity, even among persons with severe heart defects.[34-36]

Life-long, ***routine follow-up*** is considered a particularly important self-care behavior in persons with ACHD because many persons with ACHD experience a latent period between surgical correction and late complications, indicating asymptomatic disease progression.[27] Guidelines for routine follow-up are based on ACHD severity and include frequency of visits and type of provider (congenital versus general cardiologist).[27] A significant number of persons with ACHD are considered lost to follow-up. In the United States, an estimated 90% of persons with ACHD who require specialty care are not receiving care from a congenital cardiologist.[3] Even in countries with national health systems such as the Netherlands, United Kingdom, and Canada, 24-61% of persons with ACHD are no longer receiving specialty care,[37-39] although two single academic centers reported lower rates of 7%-18%.[40,41] In addition to the problem of lost to follow-up, 42-65% of persons with ACHD are reported to have significant lapses in care in the transition between pediatric and adult care.[13,42] One study found that persons with a lapse in care greater than three years were five times more likely to be symptomatic at presentation, 12 times more likely to have a new diagnosis, and three times more likely to require an urgent intervention.[13]

Cardiovascular risk behaviors including substance use (alcohol, tobacco, illicit drug), pregnancy-related behavior, weight management, and endocarditis prevention have also been reported in the ACHD population. General recommendations for ***substance use*** are the same as the acquired heart disease population including cessation of smoking and illicit drug use and no more than moderate consumption of alcohol.[11] Substance use in persons with ACHD is generally reported as equal to or lower than the general population.[16,43] Rates of current tobacco users range from 9% to 23%.[16,44-46] In young adults with moderate or severe types of ACHD, over 80% report currently drinking alcohol, 26-44% have had a binge-drinking episode in the past month, and up to 22% have used illicit drugs.[16,44] ***Endocarditis prevention*** is important for adults with cyanotic defects or mechanical devices who are at increased risk for endocarditis. Indeed, the incidence of endocarditis is estimated to be 11 per 100,000 person years in those with ACHD versus 1.5-6 per 100,000 person years in the general population.[47,48] Prophylactic antibiotics prior to dental procedures and surgery, dental hygiene, and avoiding body piercings and tattoos are important preventive behaviors. Between 25-39% of cases of endocarditis in persons with ACHD are reported as related to dental procedures or body piercing.[49-51] In young adults with ACHD, one study reported only 15% of persons saw a dentist within the past year,[44] and in the high-risk group of adults with single ventricle heart defects, 20% have been reported has having no dental visit in the past year.[16]

Maintaining a ***healthy weight*** is particularly important as persons with ACHD face additive morbidity from acquired cardiovascular conditions such as hypertension, heart failure, and coronary artery disease, and other comorbidities affecting the vascular system such as diabetes and obstructive sleep apnea.[6] Between 33-54% of persons with ACHD are reported as overweight or obese, including patients with severe ACHD.[43,45,52,53] One study found that 51% of persons with ACHD undergoing cardiac surgery were overweight or obese, had greater prevalence of hypertension, and were more likely to have postoperative renal dysfunction than person with ACHD and normal weight.[53] In adults with Tetralogy of Fallot, the most common cyanotic type of heart defect, obesity has been associated with poorer biventricular systolic function and dilation.[54] All women with ACHD are at risk for pregnancy complications associated with ACHD severity.[55] ***Contraception and pregnancy planning*** are thus important self-care behaviors in all women with ACHD. In a study examining contraceptive practices in women with a wide range of ACHD severity, 20% of women were using contraception methods contraindicated for their condition and 28% of women with high pregnancy associated risks were not using any form of contraception.[56] In pregnant women with ACHD in two single center studies, only 38-57% received care from a cardiologist during the pregnancy,[57] and 50% delivered in non-specialty ACHD centers.[58]

Antecedents of Self-Care

The FSC Framework was used to guide a review of what is currently known and identify gaps in knowledge about antecedents of ACHD self-care (Figure 1). Only 12 studies reported antecedents of individual self-care behaviors including physical activity, routine follow-up, weight management, and substance use (alcohol, tobacco, or illicit drugs) (Table 2). Individual characteristics of self-care behaviors include age, gender, living situation, distance lived from a specialty cardiac center, race, and education level. These characteristics vary by behavior and discrepant findings exist between studies. Older age has been associated with lower physical activity levels and more likelihood of having a lapse in routine follow-up,[13,19,59] but no age differences were associated with substance use.[46] Females have lower levels of physical activity.[19] Gender differences in routine follow-up and substance use are discrepant, with the majority of studies finding poorer behavior in men[38,40,45,46] and a few studies find no gender difference.[42,43] Persons with ACHD living independently from parents are more likely to have a lapse in care,[13] which may indicate the important role of family in ACHD self-care or a lack of transition of responsibility for self-care from parent to child during adolescence and young adulthood. Discrepant finding exists on the association of distance living from a specialty center with lapses in routine follow-up,[40,42] but a history of receiving care outside of a specialty center or no current medical care have been associated with lapses in routine care.[38,42] Only one study reported on race or education level, finding no association with routine follow-up behavior.[42]

Clinical characteristics related to self-care include BMI, vital signs, NYHA Class, ACHD severity, surgical history, and place of care. Lower levels of physical activity have been associated with higher BMI, higher resting systolic blood pressure, and higher NYHA Class.[17,21] A lack of association has been reported between level of physical activity and severity of heart failure (ejection fraction or BNP levels).[20] Persons with less severe types of ACHD typically have worse self-care behaviors including more lapses in routine follow-up, higher rates of obesity, and higher rates of substance use,[13,38,42,43,46] but one study reported the opposite finding.[40] Knowledge and behavioral characteristics known to be related to self-care behavior include knowledge and health beliefs. Lack of knowledge of long-term need for cardiac care have been associated with lapses in routine follow-up.[39,42] Health beliefs in regards to physical functioning have discrepant findings between individual self-care behaviors. Higher perceived physical functioning is related to higher physical activity levels,[15] but not lapses in routine follow-up.[42] In regards to the particular behavior of exercise in ACHD, two studies have shown that lower exercise self-efficacy was associated with lower levels of exercise.[21,60] No studies could be identified reporting the influence of family characteristics on self-care.

Influence of Self-Care on Health Outcomes

The FSC Framework proposes health outcomes result from self-care including proximal outcomes (symptom burden, functional and psychological health status), distal outcomes (mortality, HRU and costs, quality of life), and family outcomes (caregiver burden, caregiver health). Only 10 studies reported a relationship between individual self-care behaviors and health outcomes. The proximal outcome of functional status, including exercise capacity and symptom burden, has been related to physical activity and routine follow-up. A small to moderate, positive relationship (r=.23-.44) between physical activity level and exercise capacity has been reported in several studies.[15,18-21] Higher levels of physical activity are also associated with lower symptom burden.[34] Lapses in routine care greater than 3 years have been associated with 5 times the likelihood of being symptomatic at presentation.[13] The distal outcomes of HRU, mortality, and quality of life have been associated with physical activity, routine follow-up, and tobacco use in limited studies. One study found that persons with ACHD who had a lapse in care greater than 3 years were 3 times the likelihood to need an urgent intervention, and consumed greater health resources at that time.[13] Mortality has been associated with insufficient routine follow-up and tobacco use. In a cohort of adults with moderate severity ACHD, 48% of late deaths occurred in persons not receiving specialty care,[39] and in another study, persons with Transposition of the Great Arteries are 4.2 times the odds of death if they were a current smoker.[46] Increased quality of life has been associated with increased physical activity levels,[18,36] but not in all studies.[19,34] One study found a relationship between physical activity level and the physical component of QOL, but not the mental component.[20]

ACHD Self-Care Literature Impact

Overall, current understanding of the adequacy, antecedents, and outcomes of ACHD self-care is extremely limited (Table 3). Only 35 articles on 6 individual self-care behaviors could be identified in the literature. Of these studies, only half had sample sizes over 100 and only two-thirds included a range of heart defects versus a specific ACHD population, potentially limiting the generalizability of findings. Many of the studies were descriptive only, with only 12 (34%) and 10 (29%) of studies reporting antecedents or outcomes of self-care behaviors, respectively. In addition, there are currently no standardized self-care guidelines in the ACHD population, and thus, relatively little impact on clinical practice or patient health outcomes.

Discussion

ACHD Self-Care

Overall, a significant gap exists in even descriptively understanding self-care behaviors in ACHD. What is understood indicates that persons with ACHD may, on average, have poor self-care behaviors. No studies reporting self-care behaviors of medication adherence, diet and nutrition, immunization status, and the monitoring or management of symptoms could be identified in the literature. Further special populations of ACHD with additional self-care needs are relatively unstudied including, but not limited to, persons with heart failure who require fluid or sodium restriction, daily weights, and monitoring edema or other symptoms; persons with Fontan physiology who require further prevention of liver and renal disease; or other cyanotic groups that have air filter precautions and may have special needs related to air travel or altitude.[27] Further descriptive studies are needed to identify additional self-care behaviors that should be targeted through intervention.

Antecedents of ACHD Self-Care

Current knowledge of antecedents of ACHD self-care is even more limited than the adequacy of self-care behaviors, with only 12 studies reporting antecedents of ACHD self-care. The majority of antecedents have an inconsistent level of evidence related to only one study available or discrepant findings between studies, and no studies were identified that examined the influence of family structure, functioning, or support on adequacy of self-care (Table 3). Only severity of ACHD and ACHD knowledge had a moderate level of evidence supporting their relationship to self-care behavior, and no antecedents had a strong level of evidence. Low levels of ACHD <u>knowledge</u> among persons with ACHD are well documented and this should be further explored as an important antecedent of ACHD self-care.[15,61,62] Other unreported antecedents may also be important to ACHD self-care due to known relationships within the similar acquired heart failure (HF) population or other populations with lifelong conditions such as cystic fibrosis and sickle cell disease. <u>Individual characteristics</u> include marital status, employment status, and insurance status. In persons with acquired heart disease, those who are married, have higher levels of education and higher SES have better patterns of self-care behaviors.[63,64] <u>Clinical characteristics</u> include comorbidities and cognitive functioning. Up to 80% of persons with moderate or severe ACHD have known cognitive deficits in executive function.[65] Executive function generally refers to cognitive capacities needed for problem-solving and intentional, goal-directed behavior, which are important self-care abilities.[66] Comorbidities increase the burden of self-care through complex medication regimens, difficulty distinguishing symptoms, and disease management conflicts.[11] In the acquired HF population, comorbidities are associated with lower medication and diet adherence, difficulty interpreting healthcare professionals’ instructions for multiple illnesses, and decreased self-care confidence.[11,67,68] In the ACHD population, persons are at risk for comorbidities related to age such as diabetes and dyslipidemia or related to complications of ACHD such as pulmonary, renal, or hepatic dysfunction, and hypertension.[69]

Behavioral characteristics, including psychological and motivational factors, are well documented in the ACHD population, but very little is understood about their relationship to self-care. Approximately one-third of persons with ACHD experience depression or anxiety, although European populations report lower rates.[70] Depressive symptoms are known to reduce levels of self-care in persons with acquired HF,[11] but the relationship between self-care and depression in ACHD remains untested. Anxiety over need for re-operation, arrhythmia, or disease progression may also cripple motivation to self-care. Cardiac anxiety has been reported in ACHD. Fear of a negative evaluation is also common.[71] Health beliefs are not well understood in ACHD, but perceived physical functioning has been related to physical activity and persons with ACHD are known to have misrepresentations of their illness, such as the false belief of being "cured" which may influence self-care behaviors.

Self-efficacy is another motivating factor that is poorly understood in ACHD. Self-efficacy is a central construct in Bandura's social cognitive theory (1977) and is defined as the "conviction that one can successfully execute (a given) behavior required to produce (a given) outcome" (p. 193).[72] Self-care self-efficacy is defined as the perceived ability to successfully engage in self-care behaviors and decisions.[11] Self-care self-efficacy has been shown to be predictive of self-care behavior across multiple chronic illnesses, including the similar populations of cystic fibrosis (CF) and acquired HF.[11,73,74] In acquired HF, self-efficacy has been shown to influence self-care behaviors, and interventions improving self-efficacy have demonstrated improvements in self-care.[73,74]

Indeed, self-care self-efficacy has consistently been shown to be one of the strongest predictor of HF self-care behaviors.[75,76] It is anticipated that self-care self-efficacy was an important predictor of self-care in the ACHD population, but this remains unconfirmed. Future research addressing self-care in ACHD should include self-care self-efficacy as a predictor or antecedent of self-care.

The role of the family context is increasingly recognized as an important factor influencing level of self-care in chronic illness.[24] In acquired HF, support from family has been associated with better medication adherence and lower readmission rates,[77,78] and interventions to improve family support of self-care have been shown to improve adherence to dietary sodium restrictions.[63] Support from family, however, may also have a negative influence through critical or controlling behavior.[79] Children growing up with a chronic illness are known to experience more controlling parental behaviors than healthy subjects.[80] In studies of similar congenital illnesses, parental overprotection has lasting effects into adulthood including poorer social, psychological, and behavioral functioning.[80,81] In persons with ACHD, those who report parental overprotection from childhood are more likely to report anxiety, fear of death, and activity limitations.[82,83] In the similar adult cystic fibrosis population, qualitative studies report the importance of continued support from parents for self-care as well as the potential negative influence parents and family may have on self-care through controlling behavior.[26] This dichotomous finding is not surprising and may be similar for persons with ACHD. The relationship between parental overprotection or family support and self-care, however, has not been explored in the ACHD population. For most adults, family played the primary role in self-care for almost two decades, and maybe longer. The transition to primary responsibility for care may be thus delayed or impeded. Further, in these populations, the disease is truly a family disease, as the management of the disease has been life-long. The role of family functioning or other family factors on self-care behavior are also unknown in persons with ACHD.

Health Outcomes of ACHD Self-Care

Current knowledge of the influence of self-care on health outcomes is also very limited, with only 10 studies reporting on outcomes of individual ACHD self-care behaviors. The majority of outcomes had an inconsistent level of evidence related to only one study available or discrepant findings between studies (Table 3). Indeed, the only outcome with more than an inconsistent level of evidence was the proximal outcome of exercise capacity, which had a strong level of evidence for its relationship with daily physical activity levels. No known knowledge exists regarding the relationship between self-care family outcomes such as caregiver burden and caregiver health. In the acquired HF population, the relationship between self-care and both proximal and distal health outcomes is more fully established. Higher levels of self-care is known to improve functional capacity and symptom distress, as well as reduce hospitalizations, improve event-free survival on composite risks (death, hospitalization, emergency department visits), and quality of life.[11] Similar relationships are expected in the ACHD population, but considerable amount of work is yet to be done to determine the proximal and distal health outcomes for which self-care is a modifiable antecedent. The need to understand this relationship is critical, given the significant cardiovascular morbidity and mortality in persons with ACHD.[7,8]

Conclusions and Future Directions

Congenital heart disease is well established as a lifelong, chronic condition, but the science of self-care, a critical component of chronic illness management, remains in its infancy. This review is the first known synthesis of current knowledge regarding ACHD self-care, as well as the unique factors that may impede or nurture successful self-care and the effect of self-care on health outcomes. The results of this review highlight the paucity of ACHD self-care research, and provide a foundation to build the science of self-care in this growing population. With the enormous amount of research to be done, several key points should be made to address the current limitations and unique challenges of this population.

First, the science of self-care remains limited by a lack of self-care guidelines. Without guidelines, the ability to measure, intervene, or clinically prescribe self-care behavior in the ACHD population will remain difficult. Further, there is already some evidence that establishing guidelines can improve outcomes. In Quebec, the establishment of routine follow-up guidelines has resulted in increased referrals to congenital cardiologists and reductions in mortality.[84] Establishing comprehensive self-care guidelines, however, is complicated by the heterogenous nature of the population, and thus, the varying self-care requirements. The adult with single ventricle physiology, for instance, will have different self-care needs than the adult with repaired Tetralogy of Fallot. The needs are often categorically the same and this may be a means of standardizing, yet individualizing self-care guidelines. For example, all persons with ACHD have recommendations related to endocarditis prevention, physical activity levels, routine follow-up, and reproductive health, even though the specific recommendations may vary by type of ACHD and related complications. From a clinical perspective, the same categories of self-care requirements can by addressed with all patients even though the specific prescription will vary by person. Another option is to create separate guidelines for different major types of ACHD; this may be particularly advantageous for measurement precision in research. Whichever direction is chosen, establishing guidelines is a challenge that needs to be addressed by clinicians and researchers in the field.

The development of self-care guidelines in the acquired HF population may provide some direction for the ACHD population. In the acquired HF population, the first clinical management guidelines were published by the Agency for Health Care Policy and Research in 1994.[85] Self-care literature began to emerge concurrently, and instruments to measure self-care behaviors were first reported in the early 2000s.[86,87] With the ability to uniformly measure self-care, the acquired HF self-care literature grew exponentially and the American Heart Association and American College of Cardiology produced its first scientific statement and guidelines for HF self-care in 2009.[11] In 2007, the first ACHD clinical management guidelines were published.[27] Since that time, however, as evidenced by this literature synthesis, only a limited amount of self-care research has been conducted. Likely, this is in large part due to a lack of uniform measurement of self-care. This is a significant limitation and ought to be a priority of ACHD self-care research moving forward.

Further, there is no agreed upon definition, framework, or measurement of self-care in the ACHD population. If a unified framework can be established, the science would be more precise and generalizable across different ACHD populations and countries. Further, the science can be built more quickly, and hopefully, decrease the time from bench to bedside.

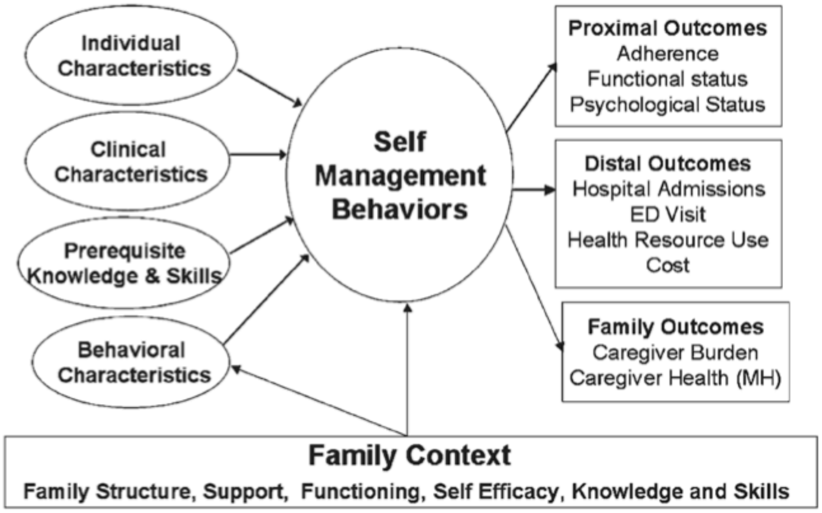
In this review, a combined definition of self-care using the FSC framework and Reigel's chronic illness theory from the acquired heart failure population was used; this may be an appropriate definition for the ACHD population by identifying separate constructs of self-care including maintenance, monitoring, and management behaviors which may differentially affect health outcomes. Multiple components of the FSC Framework appear appropriate for the ACHD population, and in particular the role of family in self-care, but further validation is required. Another key issue related to the role of the family requiring further exploration is the shift in responsibility for care from the parent to the patient as the child grows and transitions from pediatric to adult care, which may present unique challenges to self-care in this population.

It is anticipated that the establishment of a framework and measurement of self-care will lead to an exponential growth in ACHD self-care research, as in the acquired HF population, that will lead to establishing self-care guidelines.

The epidemiological landscape of congenital heart disease has shifted, and research priorities are beginning to expand in like manner to improving the quality and quantity of life in adulthood. The expansion of research priorities to self-care, however, remains limited. Every year the ACHD population grows, and the challenge of long-term management grows as well, and is becoming more complex with the now normative survival of children born with severe heart defects. The time is now to develop a better understanding of ACHD self-care so that strategies and interventions can be developed to meet and support the unique challenges faced by the ACHD population.

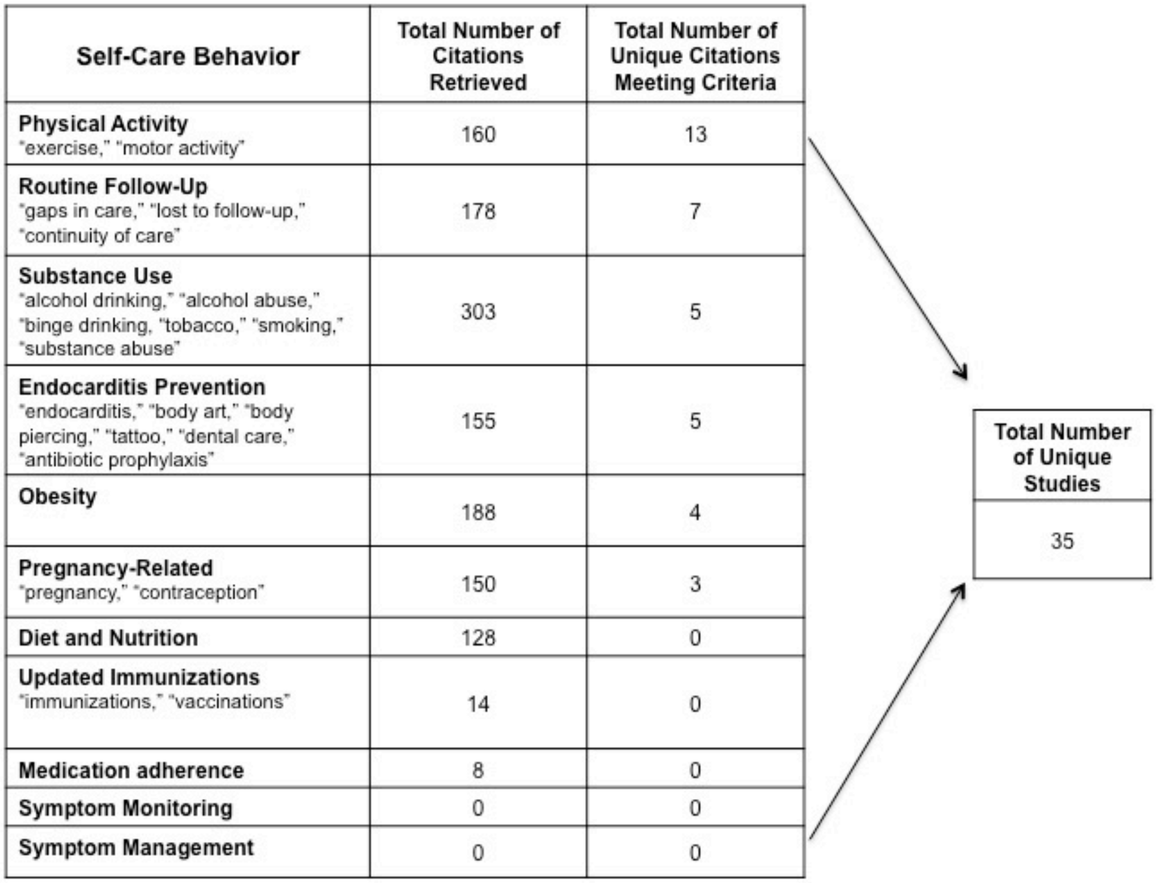
Tables and Figures

Figure 1. Family and Self-Care Framework[24]



*permission to reprint (Appendix A)

Figure 2. ACHD Self-Care Search Terms



Self-Care Behavior	Total Number of Citations Retrieved	Total Number of Unique Citations Meeting Criteria
Physical Activity "exercise," "motor activity"	160	13
Routine Follow-Up "gaps in care," "lost to follow-up," "continuity of care"	178	7
Substance Use "alcohol drinking," "alcohol abuse," "binge drinking, "tobacco," "smoking," "substance abuse"	303	5
Endocarditis Prevention "endocarditis," "body art," "body piercing," "tattoo," "dental care," "antibiotic prophylaxis"	155	5
Obesity	188	4
Pregnancy-Related "pregnancy," "contraception"	150	3
Diet and Nutrition	128	0
Updated Immunizations "immunizations," "vaccinations"	14	0
Medication adherence	8	0
Symptom Monitoring	0	0
Symptom Management	0	0

Total Number of Unique Studies
35

Table 1. Best evidence synthesis rating system used to combine the results of the studies

Strong Evidence	Consistent findings in multiple (≥2) high quality studies
Moderate Evidence	Consistent findings in one high quality study and at least one fair quality study or consistent findings in multiple fair quality studies
Inconsistent Evidence	Only one study available or inconsistent findings in multiple studies (≥2)

Table 2. Antecedents of Self-care in ACHD*

	Individual	Clinical	Knowledge & Skills	Behavioral
Physical Activity	-Female sex[19] -Older age[19,59]	-Lower BMI[15,17] -Lower systolic blood pressure[17] -Higher NYHA Class[21] -Not severity of ACHD[19] -Not ejection fraction or brain natiuretic peptide level[20]		-Health beliefs: higher perceived physical functioning [15]
Routine Follow-up	-Male sex[38,40] -Not gender, race or education level[42] -older age[13] -Not distance lived from cardiac center[40] -Distance from cardiac center[42] -Living independently from parents[13] -History of cardiac care at a non-academic center[38] or no medical care[42]	-Less severe ACHD[13,38,42] -More severe ACHD[40] -No prior heart surgery[40]	-Lack of knowledge[39,42]	-Health beliefs: Not “feeling well”[42]
Substance Use	-Male sex[45,46] -No gender differences[43] -No age difference[46]	-ACHD Severity[43,46]		
Obesity		-ACHD Severity[43]		

*Associated with higher levels of self-care behavior.

Table 3. Gaps in the Literature for ACHD Self-Care

Self-Care Behaviors	**Antecedents of Self-Care**	**Outcomes of Self-Care**
Self-care Maintenance	*Individual*	*Proximal*
Routine follow-up	**Age (I)**	**Exercise Capacity (S)**
Physical Activity	**Gender (I)**	**Symptom Burden (I)**
Smoking cessation	**Race (I)**	Psychological Status
Alcohol restriction	Marital Status	
Eat a heart-healthy diet	Employment Status	*Distal*
Endocarditis prevention:	Insurance Status	**Quality of Life (I)**
	Education Level (I)	**Mortality (I)**
Prophylactic antibiotics	**Place of Cardiac Care (I)**	**Health Resource Use (I)**
Dental hygiene		
Avoid body piercings/tattoos	*Clinical*	
	Type of ACHD	*Family*
Update immunizations	**Severity of ACHD (M)**	Caregiver Burden
Maintain a healthy weight	**Body Mass Index (I)**	Caregiver Health
Medication adherence	**Blood pressure (I)**	
Pacemaker care	Ventricular Function	
Women:	**New York Heart Class (I)**	
Appropriate birth control	Comorbidities	
Comprehensive exam prior to pregnancy	Cognitive Function	
	Knowledge/Skills	
	ACHD Knowledge (M)	
Self-care Monitoring		
Heart failure: edema checks, daily weights	*Behavioral*	
	Depression	
Change in exercise tolerance	Anxiety	
	Health beliefs (I)	
Symptom recognition	Self-efficacy	
Self-Care Management	*Family*	
Call healthcare provider	Family structure	
Take as needed medications	Family functioning	
	Family self-efficacy	
Change level of activity	Family support	

***BOLD** ≥ 1 one study on topic; (I)=Inconsistent Evidence, (M)=Moderate Evidence, (S) Strong Evidence

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CHAPTER 3: Antecedents of Self-Care in Adults with Congenital Heart Defects

Abstract

Background: Adults with congenital heart defects (ACHD) face long-term complications related to prior surgery, abnormal anatomy, and acquired cardiovascular conditions. Although self-care is an important part of chronic illness management, few studies have explored self-care in the ACHD population. The purpose of this study is to describe self-care and its antecedents in the ACHD population. **Methods:** Persons with moderate or severe ACHD (N=132) were recruited from a single ACHD center. Self-care (health maintenance behaviors, monitoring and management of symptoms), and potential antecedents including sociodemographic and clinical characteristics, ACHD knowledge, behavioral characteristics (depressive symptoms and self-efficacy), and family-related factors (parental overprotection and perceived family support) were collected via self-report and chart review. Multiple regression was used to identify antecedents of self-care maintenance, monitoring, and management. **Results:** Only 44.7%, 27.3%, and 23.3% of participants performed adequate levels of self-care maintenance, monitoring and management, respectively. In multiple regression analysis, self-efficacy, education, gender, perceived family support, and comorbidities explained 25% of the variance in self-care maintenance (R^2=.248, F(5, 123)=9.44, p<.001). Age, depressive symptoms, self-efficacy, and NYHA Class explained 23% of the variance in self-care monitoring (R^2=.232, F(2, 124)=10.66, p<.001). Self-efficacy and NYHA Class explained 9% of the variance in self-care management (R^2=.094, F(2, 80)=5.27, p=.007). **Conclusions:** Low levels of self-care are common among persons with ACHD. Multiple factors, including modifiable factors of self-efficacy, depressive symptoms, and perceived family support, are associated with self-care and should be considered in designing future interventions to improve outcomes in the ACHD population.

Introduction

Fifty years ago only one third of children born with congenital heart disease survived to 10 years of age, but today 90% of children survive to adulthood and over one million adults are living with congenital heart disease (ACHD).[1,2] Although the majority of adults had surgical repair in childhood, they are not cured. Surgical sequelae and residual defects cause late complications such as arrhythmia, heart failure, and the development of renal, pulmonary, and hepatic comorbidities.[3,4] With the exception of those with mild heart defects, complications are universal and require lifelong surveillance and illness management. Although self-care is a vital component of chronic illness management, little is understood about the practice of self-care in ACHD. A small number of studies have provided beginning evidence indicating that low levels of self-care may be a significant problem for persons with ACHD. Between 40-60% of adults, for instance, are not engaged in routine care,[5,6] and up to 85% do not meet physical activity recommendations.[7] Further, at least 20% of women do not attend pre-pregnancy counseling despite high rates of maternal and fetal complications.[5,8,9]

Lower levels of self-care are associated with poor health outcomes. In ACHD, those with a three-year lapse in care are over three times more likely to require emergent interventions, receive additional diagnoses, and experience more symptoms,[6] and those who are less physically active have greater reductions in functional capacity.[10] Thus, improving self-care may be an effective means of reducing ACHD morbidity, but the development of interventions to promote self-care is limited by the lack of evidence regarding antecedents of self-care in this population. Few have studied ACHD self-care behaviors and there is no consensus on the individual and clinical factors that may explain self-care.[11-13] Furthermore, no studies of self-care in an adult cohort of patients with congenital heart disease have examined typical antecedents important in other chronically ill populations such as knowledge, behavioral characteristics of self-efficacy or depressive symptoms, and social support.[14-16] The purpose of this study is thus to describe the practice of self-care and identify possible antecedents of self-care, so that meaningful interventions to improve self-care behaviors can be designed for the ACHD population.

Theoretical Framework

As little nursing research has been undertaken in this population, no theoretical framework has been published to guide ACHD self-care research. Given its holistic perspective and utility in the similar acquired heart failure (HF) population, this study will test Dunbar *et al's* (2008) Family and Self-Care Framework (FSCF) for use in the ACHD population (Figure 1). The FSCF was originally developed in 2008 by Dunbar *et al* from a synthesis of literature on self-care in chronic illnesses and HF, the role of family in chronic illness management, and self determination theory (SDT).[16] In this framework, self-care is defined as “activities that involve self-care decisions and actions around the recommended [HF] self-care behaviors” (p.5).[16] These activities are operationally defined according to Riegel *et al* as 1) self-care maintenance (treatment adherence activities), 2) self-care monitoring (symptom monitoring activities) and 3) self-care management (activities related to symptom response).[17] Unlike the HF population, no standard of care exists for recommended ACHD self-care behaviors. In this study, important ACHD self-care behaviors were identified from the AHA/ACC guidelines for the care of persons with ACHD and ACHD clinician expertise.[18]

The FSCF views self-care as a complex phenomenon with its success dependent on multiple antecedent domains including individual and clinical characteristics, prerequisite knowledge and skills, and behavioral characteristics such as self-efficacy, depressive symptoms, and health beliefs. Importantly, the FSCF recognizes the importance of accounting for the family context of self-care, which may directly influence self-care behaviors or indirectly influence them through behavioral characteristics. Various characteristics under each domain were tested in this study based on prior knowledge in persons with ACHD or other populations similar by disease state (acquired HF) or age and history (adults with lifelong conditions such as cystic fibrosis) (Figure 1).

Methods

A cross-sectional, descriptive study design was used to describe self-care and to determine the relationship between individual, clinical, behavioral, and family-related antecedent factors and self-care behaviors in persons with ACHD.

Sample and Setting

After receiving institutional review board approval, a convenience sample of 132 adults with moderate or severe CHD was recruited from a large Adult Congenital Heart Center clinic located in the southeastern US. Eligible adults were identified through medical chart review and recruited directly from clinic appointments.

Inclusion criteria included 1) aged 18 years and older, 2) having moderate or severe CHD per American Heart Association/American College of Cardiology guidelines,[18] and 3) English-speaking. Exclusion criteria were 1) developmental delay impairing ability to independently complete surveys, 2) adult diagnosis of ACHD, and potential confounders of a walking test including 3) current pregnancy, 4) disability preventing ability to walk, and 5) less than one month post-op from cardiac surgery. A total of 341 persons were screened for eligibility; 168 (49%) did not meet inclusion criteria with primary reasons being adult diagnosis (25.7%), developmental delay (18.3%), and having a simple heart defect (16.8%). An additional 23 (12%) declined participation. Of the 150 patients consented, 132 (88%) completed baseline data collection. Two persons (1.3%) withdrew prior to completing surveys, 1 (0.7%) was dropped due to becoming ineligible after consent, and 15 (10%) could not be reached for follow-up.

Procedures

All eligible participants were approached at clinic appointments and once informed consent was obtained, participants completed a six-minute walk test (6MWT) and received study surveys. Participants completed surveys at the clinic appointment or at home either electronically via a secure, electronic database survey system or via paper surveys returned in self-addressed, stamped envelopes. Clinical information was collected via medical chart review by the investigator. Participants were compensated with a free parking voucher and $10 gift card for their time.

Measurement

Self-Care

Self-care was measured using the Self-care Index for ACHD (SCI-ACHD). The SCI-ACHD is a revised version of the Self-Care in HF Index.[19,20] This 32-item measure of self-care was revised by the investigator with the original author's permission by adding items related to ACHD self-care including endocarditis prevention, birth control, and ACHD symptoms, and deleting some items unrelated to ACHD self-care such as salt and fluid restrictions which are not routinely prescribed except in the context of HF.[18] The SCI-ACHD includes independently scored subscales of self-care maintenance, monitoring, management (completed only by persons experiencing heart-related symptoms in past month), and self-care confidence.[17] A 4-point Likert scale is used to assess items, from 1=(Never/Not Likely/Not Confident) to 4=(Always/Very Likely/Extremely Confident).

Individual and Clinical Characteristics

Sociodemographic variables (age, gender, race, educational level, marital status, employment status, and insurance status) were collected via a standardized self-reported survey. Clinical variables (type and severity of heart defect, NYHA Class, comorbidities) were collected via medical chart review. Severity of heart defect was classified as moderate or severe based on AHA guidelines.[18] The Charlson Comorbidity Index (CCI), widely used in the acquired HF population to measure comorbidity severity, was used in this study, as no known comorbidity indices have been used for the ACHD population.[21]

ACHD Knowledge

Only one prior knowledge instrument has been developed for the ACHD population, but the instrument is long and cumbersome. Thus, the investigator-developed ACHD Knowledge Questionnaire was used in this study. This 22-item questionnaire is based on the ACHD Patient Health Passport, a 7-page booklet used to store personal health information, including health history, health risks, and self-care needs.[22] All questions are scored as correct or incorrect, summed for total score and then standardized on a 100-point scale. Higher scores reflect higher levels of knowledge.

Behavioral Characteristics

Behavioral characteristics evaluated in this study included depressive symptoms and self-care self-efficacy. Depressive symptoms was measured using the Patient Health Questionnaire-9 (PHQ-9), a widely used and well-validated 9-item, self-reported scale measuring depressive symptoms.[23] Items are scored on a 4-point Likert scale (0=not at all to 4=nearly every day) with a total score ranging from 0 to 27. Scores >10 indicate clinically relevant depressive symptoms. Internal consistency reliability is excellent in ACHD populations (CA of 0.88).[24] Self-care self-efficacy was measured using the self-care confidence subscale of the SCI-ACHD.

Family Factors

Family factors measured in this study included family self-care self-efficacy and parental overprotection. Family self-care self-efficacy was defined as the perceived support for self-care from family and was measured using the 4-item

Stanford Chronic Illness Family, Friends, and Community Self-Efficacy Scale.[25] Language related to friends and community was removed from questions so that family remained the only support mechanism, consistent with the variable to be measured. Parental overprotection was measured using the Parenting Bonding Inventory Overprotectiveness subscale,[26] a 13-item scale with excellent internal consistency reliability in the ACHD population (CA=0.89).[27] Adults are asked to rate the attitudes and behaviors of parents in the first 16 years of life. Each item is measured on a 4-point Likert scale (0=very unlikely to 3=very likely) and a total score is summed. Higher scores reflect greater overprotectiveness.

Data Analysis

Descriptive statistics were analyzed for all study variables. All instruments were scored in accordance with author guidelines and Cronbach's α was calculated for the instruments not previously tested in the ACHD population including the SCI-ACHD, CHD Knowledge Survey, and Stanford Chronic Illness Family, Friends, and Community Self-Efficacy Scale to determine reliability. Bivariate relationships between individual, clinical, knowledge, behavioral, and family antecedents and each self-care construct (maintenance, monitoring, management) were determined using Pearson's correlation. Multivariate stepwise regression analysis was used to statistically evaluate the relationship between antecedent factors and self-care. A separate analysis was run for each self-care construct. All antecedent factors (age, gender, race, education, insurance status, marital status, CHD severity, comorbidities, NYHA Class, depressive symptoms, self-care self-efficacy, parental overprotection, and perceived family support) were included in regression analysis. Factors entered the model at a .10 level of significance and stayed in the model at a .05 level of significance. Dichotomized data was used for race (white, nonwhite/other), education (high school graduate, any college or more), insurance status (private, public/uninsured), marital status (married/living with partner, widowed/divorced/single), and NYHA Class (I/II, III/IV). Multicollinearity was tested for each regression model and no confounding was present (variance inflation factor <2). All data was analyzed using SPSS version 22 and an alpha set at 0.05.

Results

Sample Characteristics

Participants were on average 36 years old with slightly more females than males (Table 1). The majority of participants were Caucasian (81%), had at least some college education, and private insurance. Approximately half of participants were married and had severe CHD, with the most common diagnoses being single ventricle physiology, Tetralogy of Fallot, and Transposition of the Great Arteries. The majority of participants had two comorbidities with the most common being heart failure, arrhythmia, and pulmonary disease. On average, participants were NYHA Class II with minimal functional limitations. Mean standardized score for CHD Knowledge was 70.7 [SD, 13.5] on a scale of 0-100, with only 8% scoring at or below the 50th percentile indicating overall average to high levels of knowledge. The majority of participants had no depressive symptoms, but 15 (11.4%) persons screened positive for major depressive disorder and were followed-up according to study protocol. On average, participants scored somewhat to very confident on their self-care self-efficacy, or perceived ability to perform self-care behaviors with a mean of 74.4 [SD, 13.9]. Participants were on average very confident in the support for self-care they received from family (Family Self-Efficacy score mean 33.2, SD 7.8), with 45 (34%) being completely confident. Mean score on the parental bonding instrument was 12.6 [SD, 7.8] with 56 (42.4%) of participant scores indicating parental overprotection in childhood.

Internal consistency reliability for the SCI-ACHD was acceptable (self-care maintenance, CA=.62; self-care monitoring, CA=.73; self-care management, CA=.60; self-care confidence, CA=.83). Reliability of the ACHD Knowledge Survey was less than adequate, CA=0.59. Low internal consistency is evidenced in other knowledge surveys in this population and may indicate low conceptual relatedness between items.[28] Reliability of the Stanford Chronic Illness Family, Friends, and Community Self-Efficacy Scale was excellent (CA=.83).

<u>Description of Self-Care</u>

Overall, persons scored a mean of 67.5 [SD, 13.7] on **self-care maintenance**, 57.2 [SD, 15.6] on **self-care monitoring**, and for the 86 persons who indicated that they experienced heart-related symptoms in the past month, a mean of 59.2 [SD, 18.7] on **self-care management**. Based on a cut-off score of 70 suggested by the developers of the original Self-Care Index for Heart Failure, less than half of the participants (44.7%) performed adequate levels of self-care maintenance, and only 27.3% and 23.3% performed adequate levels of self-care monitoring and management, respectively.[20] **Self-care maintenance** behaviors occurring on average “frequently” included avoiding tobacco, keeping routine appointments, taking medications as prescribed, avoiding excessive alcohol intake, dental hygiene, remembering to take medications, calling cardiologist prior to minor procedures, and maintaining a healthy weight. **Self-care maintenance** behaviors occurring on average only “sometimes” included taking time to relax each day, consulting with cardiologist prior to pregnancy, making healthy food choices, spending time with friends, getting adequate sleep, using a system to remember medications, using birth control, and meeting minimum exercise recommendations. All **self-care monitoring** behaviors (e.g., watching for edema, fatigue, medication side effects) occurred on average somewhat or infrequently and all **self-care management** behaviors (e.g., calling a healthcare provider, changing activity level) occurred on average somewhat or infrequently except taking an as needed medication, which occurred on average frequently.

Factors Associated with Self-Care

Individual factors significantly correlated to self-care included age, gender, education level, and insurance status (Table 2). Being older, male, having at least some college education, and having public insurance/uninsured were all moderately associated with self-care (r=.19-.37, p<.05). *Clinical factors* significantly related to self-care included NYHA Class and the Charlson Comorbidity Index. Having more comorbidities and being NYHA Class III/IV had a small to moderate association with self-care (r=.23-.30, p<.05). No statistically significant relationship was found between *knowledge* and self-care (r=.05-.06,

$p>.05$) *Behavioral factors* significantly related to self-care included both depressive symptoms and self-care self-efficacy. Lower levels of depressive symptoms and higher levels of self-care self-efficacy had a moderate relationship with self-care ($r=.24-.32$, $p<.05$). *Family factors* significantly related to self-care included perceived family support; persons with higher confidence in family support was moderately associated with self-care ($r=.18-.29$, $p<.05$).

Regression Analysis

Multiple stepwise regression was used to determine the individual, clinical, knowledge, behavioral, and family antecedent factors of self-care maintenance, self-care monitoring, and self-care management. The results of the multivariate analyses are summarized in Table 3. The final model for *self-care maintenance* included self-care self-efficacy, education level, gender, perceived family support, and the Charslon Comorbidity Index, explaining 24.8% of the variance in levels of self-care maintenance ($R^2=.248$, $F(5, 123)=9.44$, $p<.001$). Participants who were more confident in their abilities to perform self-care behaviors, had more than a high school education, were men, had higher levels of perceived family support, and a higher Charlson Comorbidity Index (indicating greater severity or number of comorbidities) had higher levels of self-care maintenance. The final model for *self-care monitoring* included age, depressive symptoms, self-care self-efficacy, and NYHA Class, which explained 23.2% of the variance in levels of self-care monitoring ($R^2=.232$, $F(2, 124)=10.66$, $p<.001$). Participants who were older, had higher levels of depressive symptoms, higher levels of confidence in abilities to perform self-care behaviors, and were NYHA Class III/IV (versus I/II) had higher levels of self-care monitoring. The final model for *self-care management* included NYHA Class and self-care self-efficacy, which explained 9.4% of the variance in levels of *self-care management* (R^2=.094, F(2, 80)=5.27, p=.007). Participants who had higher levels of confidence in abilities to perform self-care behaviors and were NYHA Class III/IV (versus I/II) had higher levels of self-care management.

Self-care self-efficacy was the only antecedent factor that significantly contributed to each construct of self-care, explaining 10%, 2.3%, and 4.2% of the variance in self-care maintenance, monitoring, and management, respectively. For self-care maintenance and management, self-care self-efficacy was the largest contributor in the model to the explanation of variance. Adequate levels of self-care maintenance, monitoring, and management (SCI-ACHD score >70) were found in 22%, 29%, and 20% of persons with the lowest tier of self-care self-efficacy versus 68%, 40%, and 39% of persons with the highest tier of self-care self-efficacy, respectively (Figure 2).

Discussion

In this study, the majority of persons with ACHD reported inadequate levels of self-care. Self-care behaviors commonly reported as performed inadequately were physical activity, pregnancy related behaviors (birth control use and pre-pregnancy planning), and stress reducing behaviors (recreation, sleep). Low levels of physical activity and pregnancy planning are consistent with the current literature.[5,7-9] Self-care behaviors of refraining from tobacco and alcohol use and practicing good dental hygiene were reported as commonly performed in this sample. This is consistent with previous studies about ACHD self-care,[29-31] and is encouraging due to this group's elevated risk for secondary hypertension, endocarditis and coronary artery disease related to aging, pulmonary and liver complications. In this study, routine follow-up was reported as generally adequate, but this is inconsistent with prior literature.[5,6] Most prior studies, however, focus on the gap in routine follow-up occurring during the transition from pediatric to adult care, whereas this study focused on routine follow-up in established ACHD patients. Thus, encouragingly, adherence to routine follow-up may improve once ACHD patients are established in specialty care.

This is the first known study to report on medication adherence and monitoring or management of symptoms. Although medication adherence is most commonly and promisingly reported by participants as adequate, less than one third of participants reported adequate monitoring and management behaviors. It is possible that through adapting to their condition for a lifetime, persons with ACHD are less likely to see a change in their condition; it may also be possible that the monitoring and management behaviors on the survey do not apply to all persons with ACHD. For instance, the behavior of "checking ankles for swelling" may only apply to a subset of participants with heart failure, and may need to be modified for future use. Overall, the results of this study indicate that self-care is an important target for intervention, especially particular self-care behaviors consistently performed at low levels in this population.

In accordance with the FSC Framework, antecedent factors of self-care in this study crossed multiple domains of characteristics. These antecedent factors of all self-care constructs (maintenance, monitoring, and management) included individual characteristics of age, gender, education level, clinical characteristics of NYHA Class and comorbidities, behavioral characteristics of depressive symptoms and self-care self-efficacy, and the family characteristic of perceived family support.

Individual Characteristics

Individual antecedents of self-care in this study included age, gender, and education level. Older age predicted higher levels of self-care monitoring, which may reflect the experience that older persons have with complications and symptoms and are thus more apt to watch for them mindfully. This has also been found in a young adult ACHD population.[13] Females had lower levels of self-care maintenance, which may be due to the low levels of pregnancy-related self-care behaviors in this population. However, this finding is consistent with other populations of chronically ill adults.[32] The relationship between lower education level and self-care is also a consistent finding across multiple chronic conditions, including adults with other lifelong conditions.[33,34] This may indicate that other factors related to but not identical to condition-specific knowledge are important for self-care in this population, including health literacy or access to health resources.

Race, marital status, and insurance status were not found to be predictors of self-care in this study. A lack of racial disparities in level of self-care is consistent with other studies on self-care behavior in this population.[11-13] Marital status and insurance status, however, are often important predictors of level of self-care in other chronically ill populations. Persons who are in supportive marriages are known to have higher levels of self-care in acquired HF.[35-37] In this study, half of participants were married, but most participants indicated that they had high levels of confidence for support from family whether they were or were not married. Adults with a lifelong condition may have more access to broader family support since their families were their primary caregivers in childhood. It is possible that this additional support may lessen the importance of marital status in this population. In other populations, lack of insurance is known to influence self-care through access to care,[38] and lack of insurance has been cited as a primary reason persons with ACHD have lapses in routine care.[39] However, no relationship was found in this study.

Clinical Characteristics

Clinical antecedents of self-care in this study included NYHA Class and Charlson Comorbidity Index, but not type or severity of heart defect. This indicates that functional severity, rather than structural severity of ACHD is most important for self-care in this population. This finding is significant because while functional severity is an important classification of disease severity in the acquired HF population, it is not commonly used in the ACHD population. Instead, structural severity of disease is the primary means by which medical professionals classify disease. This same finding was observed in a pilot study of young adults with ACHD, in which no relationship was found between structural severity based on AHA guidelines and self-care.[13] In the similar adult cystic fibrosis population, disease severity also lacks significance in relationship with self-care. Rather, treatment burden is a critical factor, highlighting the importance of functional disability as a target for self-care interventions in adults living with a lifelong condition irrespective of starting point, i.e., type of heart defect.[33,40] Future research should further explore the classification of ACHD by clinical factors other than type of heart defect.

The direction of the relationship between self-care and functional severity is an interesting but not uncommon finding in other populations. The results of this study indicate that participants who are sicker (lower NYHA Class) perform higher levels of self-care. Being sicker may indicate that a person is more experienced in dealing with his or her condition or more attuned to self-care needs due to being more symptomatic. Personal experience with ACHD may be an important factor in level of self-care, as it is in the acquired HF population.[34]

Increasing number and severity of comorbidities was significantly related to higher levels of self-care maintenance in this study. Comorbidities tend to increase the burden of self-care through complex medication regimens, difficulty distinguishing symptoms, and disease management conflicts.[34] In the acquired HF population, for instance, comorbidities are associated with lower medication and diet adherence, difficulty interpreting healthcare professionals' instructions for multiple illnesses, and decreased self-care self-efficacy.[34,41,42] In this study, the relationship was opposite. That is, persons with more comorbidities were more likely to have higher levels of self-care maintenance. Of note, persons had on average only 2 comorbidities in this study, whereas in the acquired HF population, persons commonly average five or more comorbidities.[43] A window may exist in which one or two comorbidities increases awareness and motivation to perform maintenance behaviors, but a higher number of comorbidities increases self-care complexity too much and reduces ability to perform maintenance behaviors. More objective clinical measures, such as ventricular function, may also be important antecedents of self-care, but were not explored in this study.

Knowledge

The lack of relationship between self-care and knowledge in this study is not uncommon in other chronically ill populations, although it is recognized as an important prerequisite.[34] ACHD knowledge overall was average in this study, slightly better than the low levels of knowledge previously reported in this population.[44-46] In a recent study in the acquired HF population, a lack of relationship between knowledge and self-care was explained by the interaction between knowledge and psychological distress.[47] Persons with higher levels of depression and anxiety may be less likely to recognize or respond to symptoms despite have adequate knowledge of self-care. This should be further explored in future studies of ACHD self-care.

Behavioral Characteristics

Behavior antecedents of self-care in this study included both depressive symptoms and self-care self-efficacy. Although many other behavioral characteristics exist (e.g. health beliefs and anxiety) depressive symptoms and self-efficacy were chosen to be explored in this study due to the strength of their relationship with self-care across multiple chronic conditions and the high rates of depressive symptoms reported in the ACHD population.[20,15,48,49,14] The importance of self-care self-efficacy across all domains of self-care constructs in this study is not surprising. Higher self-efficacy is commonly the strongest predictor of self-care across multiple populations living with a chronic condition including the similar adult cystic fibrosis population.[20,48,49] Two prior ACHD studies have shown that low levels of exercise were associated with lower exercise self-efficacy.[7,50] Future studies should further explore means to improve the modifiable factor of self-care self-efficacy in the ACHD population. In this study, although the finding that depressive symptoms and self-care were related as expected, the direction of the relationship was surprising and inconsistent with other populations.[34] Higher level of depressive symptoms was related to higher level of self-care monitoring. It is possible that persons with higher levels of depressive symptoms may be more vigilant in watching for changes in their symptoms, whether or not this increased attention leads to better outcomes. This surprising finding also indicates that further exploration of behavioral factors and self-care in the ACHD population is warranted.

Family Context

Family is increasingly recognized as an important factor influencing level of self-care in chronic illness, and in this study, a significant relationship was found between perceived family support and self-care maintenance even after adjusting for multiple individual, clinical, and behavioral antecedent factors.[16] Similar findings, in according with the FSCF framework, have been found in the acquired HF population. Positive support from family has been associated with better medication adherence,[51,52] and interventions to improve family support of self-care have been shown to improve adherence to dietary sodium restrictions.[53] Perceived family support should continue to be explored as a potential predictor of self-care in this population, including further knowledge as to the types of support (instrumental or emotional), or particular family relationships, that are most important for supporting self-care.

Support from family is generally positive, but it has also been shown to have a negative influence on self-care through critical or controlling behavior.[54-56] Parental overprotection refers to when these controls become unhealthy, marked by '"intrusion, excessive contact, infantilization, and prevention of independent behavior"' (p.261).[27] Children growing up with a chronic illness are known to experience more controlling parental behaviors than healthy subjects.[53] Parental overprotection has lasting effects into adulthood including poorer social, psychological, and behavioral functioning.[57,58]

In persons with ACHD, parental overprotection in childhood has been associated with symptoms of anxiety, health distress, and activity limitations.[27,59] In this study, however, parental overprotection had no relationship with self-care. It is possible that other family factors, such as current family functioning, may be more important. From a clinical perspective, family is an integrative part of ACHD care, and future research should continue to pursue which factors could be targets for self-care interventions.

Limitations

Although this study fills an important gap in understanding ACHD self-care, it is not without limitations. First, validated measures of ACHD self-care have not been previously established in the literature. Although the instrument developed by the researcher in this study is based on a well-validated instrument in acquired HF, it may require additional revisions to improve reliability and validity. The lack of explanation of variance in self-care management scores, for instance, may be related to the exclusion of important management behaviors in this population versus the acquired HF population, rather than a lack of applicable antecedents. Further, recent evidence supports a multidimensionality of the self-care maintenance and management subscales that significantly improve reliability.[60] Future studies with a larger sample should investigate the dimensions of the SCI-ACHD. Further, a lack of standard of care for self-care behaviors in the ACHD population may significantly contribute to lower reported levels of self-care. Second, cross-sectional and descriptive design does not allow for the determination of cause and effect relationships. This study did, however, allow for the exploration of many potential antecedents of self-care to generate future hypotheses and guide intervention development. Also, participants were recruited from one ACHD center, potentially limiting generalizability of findings and causing selection bias as adults followed in non-specialty centers or those not currently engaged in routine care (as high as 60% in the United States[39]) were not included. The importance of antecedent factors of self-care may vary in these groups. Finally, self-care was measured via self-reported surveys and may not reflect actual behavior. Although this may positively skew results, we did observe significant variability in the reported self-care behaviors that supports integrity of self-care behavior reporting in this population.

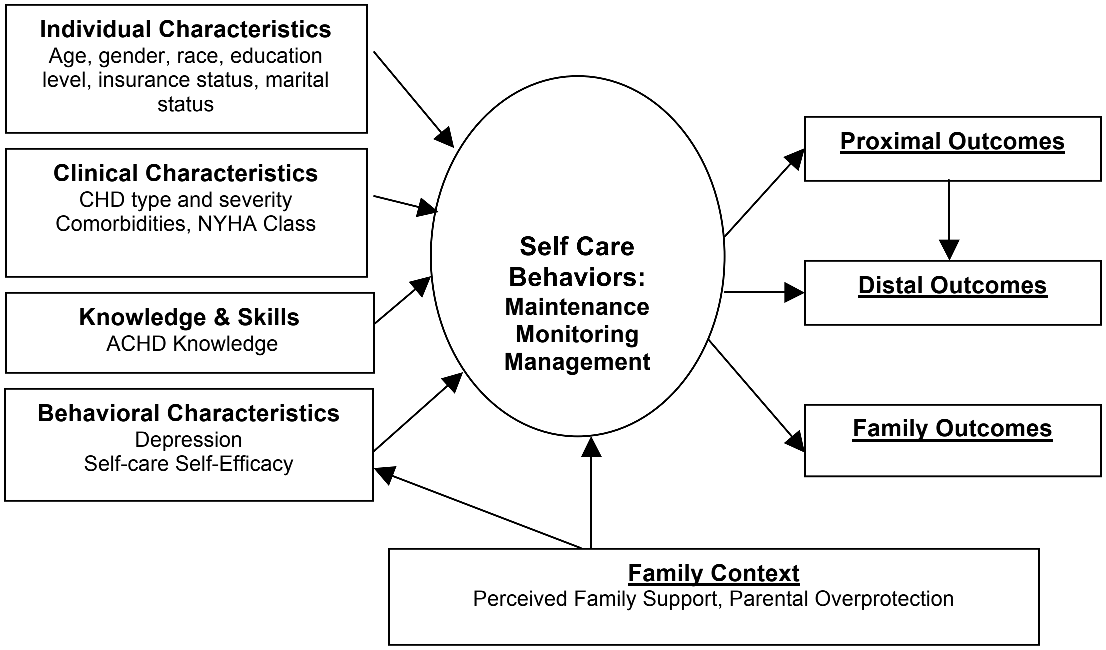
Conclusions

The distribution of congenital heart disease has changed dramatically over the last four decades, with the burden of disease shifting from the acute, life-threatening illness of the newborn to the chronic, life-limiting illness of the adult.[2] Evidence-based treatment and management of ACHD remains in its early stages, with the first AHA/ACC guidelines published just six years ago.[18] The role of self-care, a vital component of chronic illness management, also remains evidence-scarce in this population.

Overall, this study indicates that a large portion of persons with ACHD do not have adequate levels of self-care. Certain behaviors may be particularly important for improvement including physical activity, and pregnancy-related behaviors among women with ACHD. Although some predictors of self-care in this study were nonmodifiable (age, gender, education level), these factors may help to identify persons at risk of low levels of self-care that may benefit most from intervention. Self-care self-efficacy shows promise as a modiable risk factor for future interventions, as well as family support. The positive relationship between depressive symptoms and self-care was unexpected and should also be further explored in future studies. Another important finding in this study was difference between self-care maintenance, monitoring, and management. Different factors best explain these constructs, indicating both the complexity of self-care and the importance of targeting different subpopulations or different interventions based on the aspect of self-care that is trying to be improved. Although multiple factors were identified as important for self-care, there may be other factors not represented in this study that may add to explanation of variance in self-care. Factors important in other populations include cognition, sleep, health literacy, patient-physician interactions, and general social support.[34] Future research is needed in to shape guidelines and policies to support ACHD self-care. This study is an important first step in expanding the evidence-base for the specific antecedents associated with self-care in ACHD that will lead to the development of evidence-based interventions supporting self-care, and ultimately, the reduction both the personal and societal burden of ACHD.

Tables and Figures

Figure 1. Family and Self-Care Framework for ACHD*



**Adapted from the Family and Self-Care Framework for HF*[16]

Table 1. Individual and Clinical Characteristics

Variable	**Mean [SD]/N (%)**
Age	35.8 [11.4]
Gender	
Male	59 (44.7)
Female	73 (55.3)
Race	
White	107 (81.1)
Black	18 (13.6)
American Indian/Alaskan	1 (0.8)
Asian	4 (3.0)
>1 Race	2 (1.5)
Martial Status	
Single	57 (43.2)
Married	63 (47.7)
Living with Partner	5 (3.8)
Divorced	6 (4.5)
Widowed	1 (0.8)
Insurance	
Private	81 (61.4)
Medicare/Medicaid	43 (32.6)
Uninsured	8 (6.1)
Education	
< High school	2 (1.5)
High school	59 (44.7)
College degree	50 (37.9)
Graduate degree	21 (15.9)
Heart Defect Severity	
Moderate	61 (46.2)
Severe	71 (53.8)
NYHA Class	
I	22 (16.7)
II	73 (55.3)
III	36 (27.3)
IV	1 (0.8)
Charlson Comorbidity Index	1.61 [1.66]

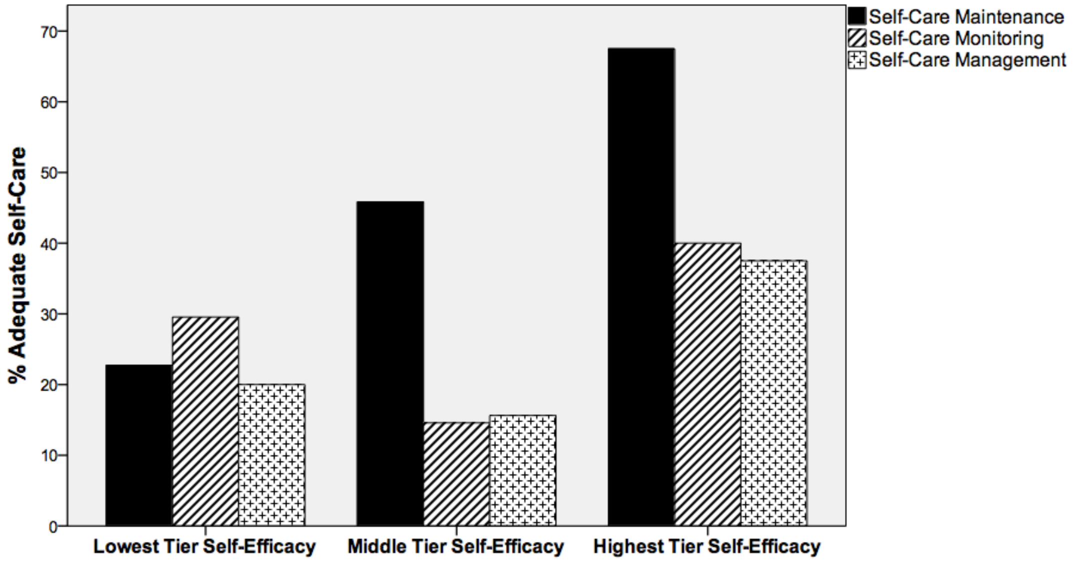
Table 2. Bivariate Relationship Between Self-Care and Antecedent Factors

Antecedent Factor	*Self-Care Maintenance (r, p-value)*	*Self-Care Monitoring (r, p-value)*	*Self-Care Management (r, p-value)*
Age	-0.05, .57	**0.37, <.001**	0.08, .45
Gender	**0.27, .002**	-0.14, .10	-0.07, .51
Race	0.08, .39	0.06, .50	-0.10, .38
Education Level	**0.32, <.001**	**-0.19, .03**	-0.07, .54
Insurance Status	0.16, .07	**-0.22, .01**	-0.05, .68
Marital Status	0.04, .68	0.10, .27	-0.12, .26
Severity Heart Defect	0.10, .25	0.07, .41	-0.03, .77
NYHA Class	-0.14, .10	**0.30, .001**	0.18, .09
Charlson Index	0.13, .13	**0.23, .008**	0.15, .17
ACHD Knowledge	0.06, .50	-0.05, .59	-0.05, .66
Depressive Symptoms	**-0.29, .001**	**0.24, .005**	-0.05, .65
Self-Care Self-Efficacy	**0.32, <.001**	0.11, .21	0.20, .07
Parental Overprotection	**-0.18, .04**	-0.06, .50	0.01, .93
Perceived Family Support	**0.29, .001**	-0.10, .24	-0.01, .91

Table 3. Multiple Regression Analysis

	Self-Care Maintenance			
	β	*SE*	*p-value*	*Model Statistics*
Self-Care Maintenance				Model F=9.44; R^2=.248 p<.001
Self-care Self-efficacy	1.13	.33	.001	
≤Highschool Education	-6.22	2.25	.007	
Male Gender	4.94	2.21	.027	
Perceived Family Support	0.31	0.14	.032	
Charlson Comorbidity Index	1.32	0.66	.046	
Self-Care Monitoring				Model F=10.66;R^2=.232 p<.001
Age	0.10	0.03	<.001	
Depressive Symptoms	0.21	0.07	.004	
Self-care Self-efficacy	0.27	0.09	.004	
NYHA Class III/IV	1.62	0.71	.025	
Self-Care Management				Model F=5.27; R^2=.094 p=.007
Self-care Self-efficacy	0.21	0.08	.008	
NYHA Class III/IV	1.36	0.53	.011	

Figure 2. Relationship Between Adequate Level of Self-care and Self-efficacy



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CHAPTER 4: Relationship Between Self-Care, Functional Health, and Health Resource Use in Adults with Congenital Heart Defects

Abstract

Background: Many adults with congenital heart disease (ACHD) face progressive functional decline and increasing health resource use (HRU) related to long-term complications. The ability of ACHD self-care to influence functional decline is relatively unknown. **Objective:** The purpose of this study was to determine the relationship between self-care (maintenance, monitoring, and management behaviors), functional health (exercise capacity, symptom burden) and HRU. **Methods:** Adults with moderate or severe ACHD (mean age 36 ± 11 years, 55% female, 81% Caucasian) were recruited from a single ACHD center (N=132). Participants completed baseline surveys on self-care, symptom burden, and sociodemographics, and a six-minute walk test (6MWT). Clinical data were collected by chart review. Self-reported HRU was collected prospectively for 3 months. Bivariate correlations, linear and logistic regression analyses were used. **Results:** Mean total distance walked on the 6MWT was 1365 ± 275 feet. Participants experienced 7.4 ± 4.8 symptoms in the previous month. A total of 101 (80.8%) participants experienced 310 healthcare encounters (275 office visits, 21 hospitalizations, 20 ED visits); 73 persons (58.4%) had > 1 HRU encounter. In multivariate analysis, self-care management ($\beta=-44.20$, $p=.001$) and monitoring ($\beta=-14.59$, $p=.03$) were independently associated with the 6MWT; self-care maintenance and monitoring were independently associated with symptom burden ($\beta=-.39$, $p<.001$; $\beta=1.52$, p<.001); higher self-care monitoring was an independent predictor of ≥ 1 HRU encounter (OR=1.21, 95% CI [1.09, 1.35], p<.001). **Conclusions:** Self-care may be an important, modifiable predictor of functional health and HRU in the ACHD population. The development of interventions to improve self-care should be pursued in future studies.

Introduction

Congenital heart disease is one of the most successfully treated forms of heart disease. Childhood survival rates have changed from less than 50% to 90% in just four decades.[1] The success of pediatric treatment, however, has led to an emerging population of chronically ill adults. With the exception of persons with simple defects, all adults with congenital heart disease (ACHD) face complications from surgical sequelae and residual defects such as heart failure, arrhythmia, and multi-organ comorbidities, often requiring reintervention or reoperation.[2,3,4] Many adults face progressive functional decline with increasing exercise intolerance and clinical symptoms, and a related increase in health resource use (HRU). Indeed, between 1998 and 2005, the number of ACHD hospitalizations in the United States doubled, growing from 36,000 to 72,000 admissions. The total estimated ACHD healthcare spending increased from $691 million to $3.16 billion,[5] and the most recent data from 2007 show an additional 10% increase in hospitalizations.[6] HRU is only expected to increase with the 5% annual growth in the ACHD population and the now normative survival of children with severe defects.[7]

Although persons with ACHD are born with their condition, not all long-term complications are inevitable. Limited evidence suggests that self-care, or the behaviors and decisions a person makes to maintain health as well as monitor and manage symptoms, may influence the development and timing of complications.[8] Persons with ACHD who have lower physical activity levels, for instance, are known to have greater reductions in exercise capacity.[9] Exercise capacity is among the few known predictors of worse health outcomes in ACHD, including hospitalizations, transplant, and death.[10,11] A lack of routine follow-up has also been shown to lead to emergent interventions, additional diagnoses, and a higher number of symptoms.[12] This limited evidence, in addition to the known relationship between self-care and health outcomes in the similar acquired heart failure (HF) population,[1f3] suggests that self-care may be an important, modifiable predictor of functional decline persons with ACHD. Thus far, however, the evidence is limited to specific individual behaviors, and no evidence exists on the importance self-care behaviors such as medication adherence or monitoring and managing of symptoms. If ACHD self-care significantly contributes to poorer health outcomes, interventions may be targeted at improving self-care in order to reduce poor outcomes in this growing population. Therefore, the purpose of this study is to determine the relationship between ACHD self-care and functional health (exercise capacity and symptom burden) and HRU (office visits, emergency department (ED) visits, and hospitalizations). We hypothesized that higher levels of self-care would be related to increased exercise capacity, lower symptom burden, and less HRU.

Methods

A cross-sectional, descriptive design was used to examine the relationship between self-care, functional capacity, and symptom burden, and a prospective, descriptive component was used to examine the relationship between self-care and HRU.

Sample and Setting

After receiving IRB approval, a convenience sample of 132 adults with moderate or severe ACHD was recruited from the Emory Adult Congenital Heart Center clinic. Eligible adults were identified through chart review and recruited directly from clinic appointments. Inclusion and exclusion criteria are provided in Table 1. Eligible participants were approached at clinic appointments and once informed consent was obtained, participants completed a walking test and study surveys. HRU data were collected monthly by electronic survey or phone. Clinical data were collected by chart review.

Measurement

Self-Care

Self-care was operationalized as three separate constructs: self-care maintenance (behaviors that maintain physiologic stability), self-care monitoring (attention to changes in symptoms), and self-care management (response to symptoms when they occur). Self-care was measured by the 32-item Self-care Index for ACHD (SCI-ACHD), a modified version (with the original author's persmission) of the Self-Care in HF Index[14,15] by adding or removing symptoms as relevant to ACHD.[38] Independently scored subscales of self-care maintenance, monitoring, and management (only for persons experiencing heart-related symptoms in past month), using a 4-point Likert scale. Scores were standardized on a 100-point scale with higher scores reflecting higher levels of self-care.

Exercise Capacity

Exercise capacity was measured using the Six Minute Walk Test (6WMT), a widely used objective measure of exercise capacity in acquired HF with moderate to strong correlations with cardiopulmonary exercise testing (CPET).[16,17] The test was given in accordance with American Thoracic Society guidelines,[21] with participants asked to walk as far as possible along a level, marked hallway for six minutes. Total distance walked in feet was used in analysis.

Symptom Burden

Symptoms were defined as an individual's subjective experiences of changes in their physical or mental state.[22] Symptom burden was defined as the average perceived frequency, severity, and affect on life enjoyment of symptoms.[23,24] Symptom burden was measured using the Congenital Heart Disease Symptom Survey (CHDSS), a revision of the Heart Failure Symptom Survey (with the original author's permission) by adding or removing symptoms as relevant to ACHD.[24] Persons rate the frequency, severity, and affect on enjoyment of life of 23 symptoms over a one-month timeframe, using a 5-point Likert Scale. The average score of frequency, severity, and affect on enjoyment of life were used to create a symptom burden score for each symptom, and summed for an overall symptom burden.

Health Resource Use

HRU, defined as total number of office visits, ED visits without hospitalization, and hospitalizations within 3 months of recruitment, was measured using a self-reported survey. All hospitalizations were reported but only unplanned hospitalizations were used in analysis. Hospitalizations and ED visits were categorized as cardiac versus non-cardiac based on admitting diagnosis or self-report. HRU occurring at the primary facility was validated by chart review, and less than 15% of HRU was underreported.

Demographic and Clinical Variables

Sociodemographic variables (age, gender, race, educational level, marital status, employment status, and insurance status) were collected via a standardized self-reported survey. Clinical variables (Charlson Comorbidity Index, type and severity of heart defect, New York Heart Association (NYHA) Class) were collected via medical chart review. Severity of heart defect was classified as moderate or severe based on AHA guidelines.[38] The Charlson Comorbidity Index (CCI), widely used in the acquired HF population to measure comorbidity severity, was used in this study, as no known comorbidity indices have been used for the ACHD population.[25]

Data Analysis

All instruments were scored in accordance with author guidelines and Cronbach's α was calculated for instruments not previously tested in the ACHD population including the SCI-ACHD and CHDSS. Descriptive statistics were analyzed for all study variables. Bivariate relationships between level of self-care and functional health (exercise capacity and symptom burden) were determined using Pearson's correlation. Multivariate stepwise regression analysis was used to statistically evaluate the relationship between level of self-care maintenance, monitoring, and management, and functional health outcomes, controlling for clinical (severity of ACHD and CCI) and sociodemographic (age, gender, race) factors. Total HRU was positively skewed, with approximately half of participants (55.2%) having ≤ 1 HRU encounter. Thus, HRU was dichotomized as ≤ 1 or >1 encounter. To determine if self-care was an independent predictor of 3-month HRU, univariate and multivariate logistic regression with forward likelihood ratio variable selection was used, controlling for clinical factors (severity of ACHD, CCI, NYHA Class) and additional sociodemographic factors (age, gender, race, education level, marital status, employment, and insurance status) to control for the resources and support for utilizing healthcare. All data was analyzed using SPSS version 22 and an alpha set at 0.05.

Results

A total of 341 persons were screened for eligibility; 168 (49%) did not meet inclusion criteria [adult diagnosis (25.7%), developmental delay (18.3%), simple ACHD (16.8%)]’ 23 persons (12%) declined participation. Of the 150 patients recruited, 132 (88%) completed baseline data collection; 2 (1.3%) withdrew prior to completing surveys, 1 (0.7%) was dropped due to not meeting inclusion criteria, and 15 (10%) could not be reached for follow-up. A total of 125 (95%) completed 3-month follow-up for HRU.

Sample Characteristics

Participants were on average 36 years old with slightly more females than males. The majority were Caucasian, married, with private insurance, and a highschool degree (Table 2). Approximately half of participants severe ACHD with the most common diagnoses being single ventricle physiology, Tetralogy of Fallot, and

Transposition of the Great Arteries. The majority had two comorbidities; the most common were heart failure, arrhythmia, and pulmonary disease. The majority of participants were functional NYHA Class II, indicating minimal daily limitations. Internal consistency reliability for the SCI-ACHD was acceptable (self-care maintenance, CA=.62; self-care monitoring, CA=.73; self-care management, CA=.60). Reliability of the CHDSS was excellent (frequency, CA=.86; severity, CA=.87; enjoyment, CA=.89.)

Self-Care

On average, persons scored 67.5 [SD, 13.7] on self-care maintenance, 57.2 [SD, 15.6] on self-care monitoring, and for the 86 (65.1%) persons who indicated that they experienced heart-related symptoms in the past month, 59.2 [SD, 18.7] on self-care management. Based on a cut-off score of 70 suggested by the developers of the original Self-Care Index for Heart Failure, less than half of the participants (44.7%) performed adequate levels of self-care maintenance, and only 27.3% and 23.3% performed adequate levels of self-care monitoring and management, respectively.[15]

Health Outcomes

Exercise Capacity

Mean total distance walked on the six-minute walk test was 1365 ± 275 feet. In bivariate analysis, a moderate, negative relationship was found between total distance walked and self-care monitoring (r=-.28, p=.001) and self-care management (r=-.35, p=.001), but no relationship was found with self-care maintenance (r=.08, p=.38). Participants who walked farther had lower scores on self-care monitoring and management. After adjusting for sociodemographic and clinical factors (age, gender, race, severity of CHD, Charlson Comorbidity Index), self-care management (β=-44.20, p=.001) and self-care monitoring (β=-14.59, p=.03) remained significant predictors of total distance walked.

Symptom Burden

Participants experienced an average of 7.4 ± 4.8 symptoms in the previous month. The most frequently occurring symptoms included fatigue, headache, palpitations, anxiety, shortness of breath performing usual activities, and difficulty sleeping. Overall symptom burden was 17.9 ± 14.0 (range 1.3-64.0). The most burdensome symptoms included problems with sexual functioning, difficulty climbing stairs, difficulty sleeping, fatigue, excessive sweating, and abdominal pain. In bivariate analysis, overall symptom burden was significantly and negatively related to self-care maintenance (r=-.29, p=.001), and positively to self-care monitoring (r=.40, p=<.001) and self-care management (r=.25, p=.02). After adjusting for clinical and sociodemographic factors (age, gender, race, CHD severity, Charlson comorbidity index), only self-care maintenance and self-care monitoring were independent predictors of overall symptom burden (β=-.39, p<.001; β=1.52, p<.001, respectively). The relationship between the top ten most burdensome symptoms and self-care maintenance and monitoring are depicted in Figure 1.

Health Resource Use

A total of 102 (81.6%) participants experienced 310 encounters with the healthcare system (Table 3). 73 (58.4%) participants experienced > 1 HRU encounter. The majority of the encounters were office visits, averaging 2.2 [SD, 2.1]. Nine planned hospitalizations, 12 unplanned hospitalizations, and 20 ED visits occurred. All planned hospitalizations were cardiac surgery or related procedures. Most unplanned hospitalizations (58.3%), but only a minority of ED visits (15.0%) were cardiac-related. Self-reported reasons for the majority of ED visits were abdominal pain, migraines, and post-procedural issues. A minority of hospitalizations (23.8%), but the majority of ED visits occurred at non-ACHD centers (65.0%).

In univariate analysis, self-care monitoring was significantly related to having > 1 HRU encounter (OR=1.21, 95% CI [1.09, 1.35], p<.001). Self-care maintenance (OR=1.00, 95% CI [.98, 1.03], p=.63) and self-care management (OR=1.21, 95% CI [.97, 1.51], p=.09) were not significantly related HRU. After adjusting for sociodemographic and clinical variables, self-care monitoring remained significantly related to having > 1 HRU encounter and in fact, was the only variable to enter the model (OR=1.21, 95% CI [1.09, 1.35], p<.001). For every unit increase in the self-care monitoring score (indicating better self-care monitoring), the likelihood of having more than > 1 HRU encounter increased by 21%.

Discussion

This study represents the first known study to examine the relationship between self-care and functional health (exercise capacity and symptom burden) and HRU in the ACHD population. After adjusting for clinical and sociodemographic factors, self-care was independently associated with exercise capacity and symptom burden and was an independent predictor of 3-month HRU.

Exercise Capacity

In this study, mean distance walked on the 6MWT was significantly greater than the validated 984-foot cut-point for poorer outcomes in the acquired HF population.[26] A higher cut-point of 1476 feet has been suggested for the ACHD population, but this has not been validated.[27] In this study, self-care monitoring and self-care management were independently associated with total distance walked on the 6MWT. The direction of the relationship, however, was unexpected. As self-care monitoring and management scores increased, total distance walked decreased. Thus, persons with more adequate self-care monitoring and management had lower exercise capacity. At face value, this could indicate that better self-care was associated with worse outcomes. While possible, this finding more likely reflects that persons with higher functional limitations are more likely to perform behaviors to monitor and manage symptoms simply because they have more symptoms. Other studies aiming to determine the relationship between self-care and exercise capacity are limited to the self-care maintenance behavior of physical activity. Daily physical activity has been moderately associated with exercise capacity (r=.40), and some evidence indicates that increasing physical activity can increase exercise capacity in persons with ACHD.[28,29] Certain medications such as ACE inhibitors and pulmonary vasodilators have shown promise to increase exercise capacity in subpopulations of persons with ACHD, but data are limited and the relative contribution of the self-care maintenance behavior of medication adherence is unknown.[2]

Symptom Burden

In this study, self-care maintenance and self-care monitoring were independently associated with overall symptom burden, explaining 28% of its variance. The relationship between self-care maintenance and symptom burden was expected. Higher symptom burden was associated with less adequate self-care maintenance and explained 12% of the variance in symptom burden scores. The relationship between self-care monitoring and symptom burden, however, was unexpected. Higher symptom burden was associated with more adequate self-care monitoring, and explained 16% of variance in symptom burden scores. This finding is similar to the relationship between self-care monitoring and exercise capacity, and again, most likely, suggest that persons with higher symptom burden are more likely to perform behaviors to monitor symptoms simply because they have more symptoms.

No other known studies have examined the relationship between symptom burden and self-care in the ACHD population to compare these findings. Indeed, very few studies have even examined the symptom experience of persons with ACHD.[30] In the acquired HF population that experiences similar symptoms, symptom burden is known to increase with poor self-care maintenance behaviors such as a lack of adherence to sodium or fluid restrictions.[23,31] Self-care maintenance may be an important area of intervention to reduce symptom burden in the ACHD population and should be further explored with a more in-depth study of specific self-care behaviors.

Health Resource Utilization (HRU)

In this study, self-care monitoring was an independent predictor of HRU in the ACHD population. Again, similar to its relationship with exercise capacity and symptom burden, the direction of the relationship was unexpected. Persons with better self-care monitoring had greater HRU. This may in fact be due to early detections of symptoms and changes that prompted seeking attention. Indeed, it is possible that increased HRU is protective of adverse health outcomes. The majority of HRU in this population was office visits. Persons with ACHD who attend routine visits or follow-up more frequently may be less likely to have adverse health outcomes. Other studies have shown that gaps in routine care increase the need for emergent interventions and increase symptom burden in the ACHD population.[12] Future studies should explore whether HRU has a positive or negative effect on adverse outcomes in the ACHD population. Even a cardiac hospitalization in this population could be evidence of excellent self-care, in that reoperations are expected in certain subsets of persons with ACHD, and when performed in a timely manner, reduce the risk of long-term morbidity. In this study, seven (28%) of hospitalizations were for cardiac surgery or pacemaker replacements; it is unknown whether these surgeries could be considered timely or preventative of poorer future outcomes. It does highlight, however, the unique issues faced by persons with ACHD in regards to HRU that may differ from the acquired HF population and should be taken into consideration in future studies exploring HRU.

Limitations

Although this study fills an important gap in understanding the relationship between ACHD self-care and health outcomes, it is not without limitations. First, validated measures of self-care and symptom burden in ACHD have not been established in the literature; although the self-care and symptom burden instruments developed by the researcher in this study are based on well-validated instruments in acquired HF, they may require additional revisions to improve reliability and validity, and to validate cut points for adequate self-care. Also, measures of self-care were self-reported and do not reflect actual behavior, which may result in biased results. Second, the cross-sectional and descriptive component of this study does not allow for the determination of cause and effect relationships between self-care and functional health outcomes, but these findings confirm the potential of self-care to be a modifiable predictor of poorer outcomes, helping to generate future hypotheses and guide intervention development. Lastly, participants were recruited from one ACHD center, potentially limiting generalizability of findings and causing selection bias as many people with ACHD are followed in non-specialty centers.

Conclusions

With the increasing number of persons living with ACHD and particularly the recent growth in persons with severe ACHD, the need to develop interventions to reduce morbidities associated with long-term complications is critical. Few prior studies have examined the relationship between self-care and health outcomes, despite the importance of self-care in chronic illness management. This study represents an important shift in prioritizing the chronic management of CHD, paralleling the shift in the burden of CHD from childhood mortality to adult morbidity that has already occurred. The results of this study indicate that self-care may be an important, modifiable predictor of reduced functional health and increasing HRU in the ACHD population. In the similar acquired HF population, interventions to improve self-care have been shown to improve outcomes. Future research is needed to further explore the role of self-care in health outcomes in the ACHD population, including the development and testing of interventions to improve self-care.

Tables and Figures

Table 1. Inclusion and Exclusion Criteria

Inclusion Criteria	**Exclusion Criteria**
≥ 18 years	Developmental delay impairing ability to independently complete surveys
Moderate or severe ACHD per American Heart Association/American College of Cardiology guidelines	Adult diagnosis, and potential confounders of a walking test including current pregnancy, physical disability, less than one month post-op from cardiac surgery
English-speaking	

Table 2. Baseline Characteristics

Variable	**Mean [SD]/N (%)**
Age	35.8 [11.4]
<u>Gender</u>	
Male	59 (44.7)
Female	73 (55.3)
<u>Race</u>	
White	107 (81.1)
Black	18 (13.6)
American Indian/Alaskan	1 (0.8)
Asian	4 (3.0)
>1 Race	2 (1.5)
<u>Martial Status</u>	
Single	57 (43.2)
Married	63 (47.7)
Living with Partner	5 (3.8)
Divorced	6 (4.5)
Widowed	1 (0.8)
<u>Insurance</u>	
Private	81 (61.4)
Medicare/Medicaid	43 (32.6)
Uninsured	8 (6.1)
<u>Education</u>	
< High school	2 (1.5)
High school	59 (44.7)
College degree	50 (37.9)
Graduate degree	21 (15.9)
<u>Heart Defect Severity</u>	
Moderate	61 (46.2)
Severe	71 (53.8)
<u>NYHA Class</u>	
I	22 (16.7)
II	73 (55.3)
III	36 (27.3)
IV	1 (0.8)
Charlson Comorbidity Index	1.61 [1.66]

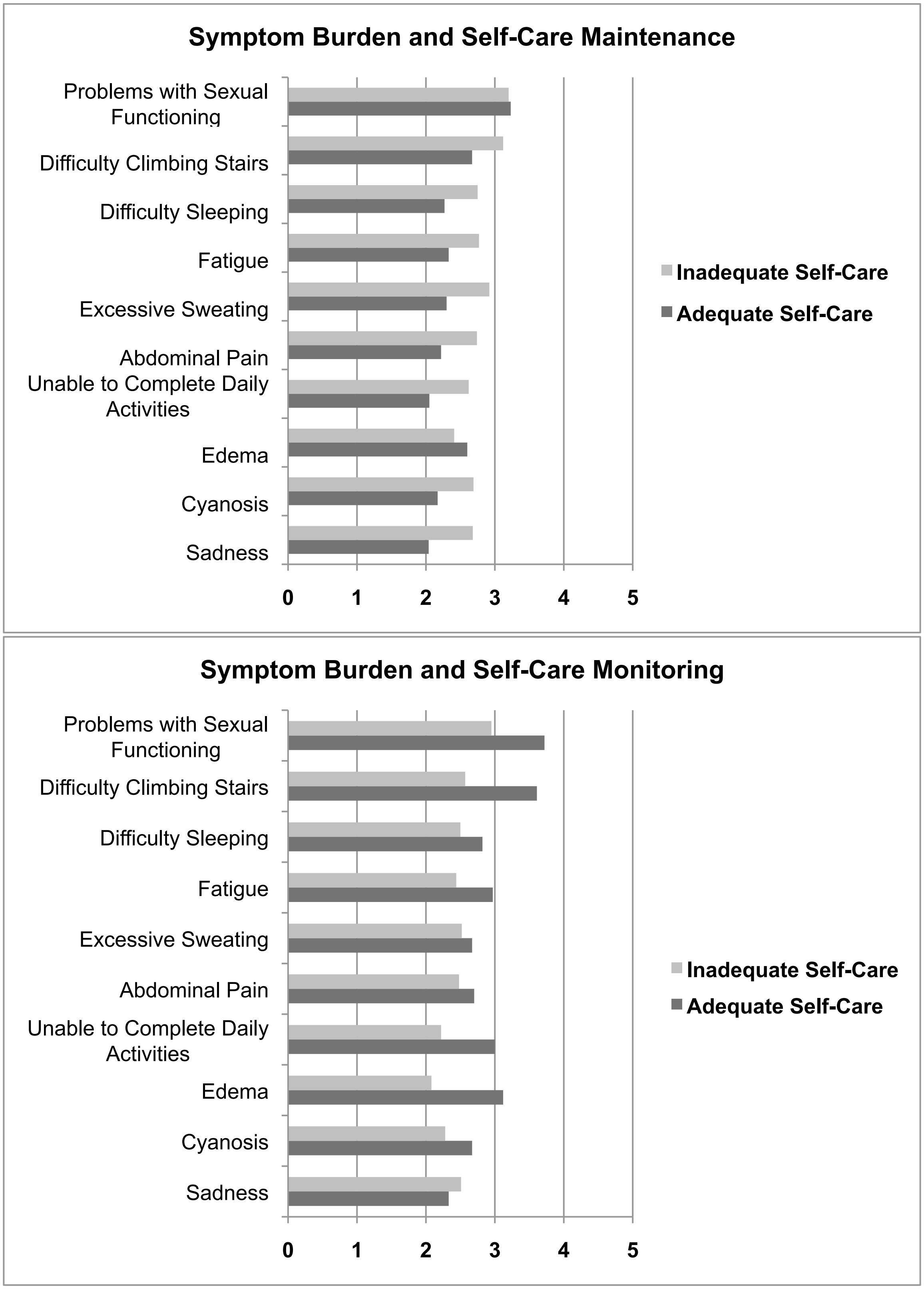
Table 3. Health Resource Use (N=125)

Variable	**Mean[SD]/N (%)**
Total Health Resource Use	2.4 [2.4] (range 0-10)
Total Number Office Visits	2.2 [2.1]
Type Number of Office Visits	275
Cardiology	124 (45.1%)
Non-Cardiology	151 (54.9%)
Number of Hospitalizations	21
Planned cardiac-related	9 (42.9%)
Unplanned	12 (57.1%)
Cardiac-related	7 (58.3%)
Non-cardiac related	5 (41.7%)
Non-ACHD specialty center	5 (23.8%)
Number of Emergency Visits	20
Cardiac-related	3 (15.0%)
Non-cardiac related	17 (85.0%)
Non-ACHD specialty center	13 (65.0%)

Table 4. Multivariable Regression Analysis

Health Outcomes	Model Statistics			
	β	*SE*	*p-value*	
Functional Capacity (All Subjects)				Model F=8.45; R^2=.10 p<.001
Age	-5.28	2.17	.02	
Self-care Monitoring	-14.59	6.56	.03	
Functional Capacity (Subjects filling out self-care management subscale)				Model F=11.2; R^2=.11 p=.001
Self-care Management	-44.20	13.2	.001	
Overall Symptom Burden (All Subjects)				Model F=21.09;R^2=.32 p<.001
Self-care Monitoring	1.52	.29	<.001	
Self-care Maintenance	-.39	.08	<.001	
Comorbidity Index	2.06	.63	.001	
Overall Symptom Burden (Subjects filling out self-care management subscale)				Model F=5.06;R^2=.16 p=.003
Self-care Management	1.28	.68	.06	
Comorbidity Index	2.11	.84	.01	
Male Gender	-6.66	3.10	.03	

Figure 1. Relationship Between Most Burdensome Symptoms and Self-Care



*Symptom Burden=Mean of Symptom Frequency, Severity, and Affect on Life Enjoyment (5-pt Likert Scale)

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CHAPTER 5: Conclusion

The purpose of this dissertation study was to examine antecedents and outcomes of self-care in adults with congenital heart disease (ACHD), conceptually guided by the Family and Self-Care (FSC) Framework. In this framework, self-care is defined as “activities that involve self-care decisions and actions around the recommended [heart failure] self-care behaviors” (p.5).[1] These self-care activities were operationally defined in this dissertation research according to Riegel *et al* as 1) self-care maintenance (treatment adherence activities), 2) self-care monitoring (symptom monitoring activities) and 3) self-care management (activities related to symptom response).[2] The first aim of this study was to examine the relationship between individual, clinical, knowledge, behavioral, and family antecedent factors and level of self-care. The second aim was to determine the relationship between level of self-care and health outcomes of functional capacity, symptom burden, and health resource use (HRU).

A cross-sectional, descriptive design was used to examine the relationship between antecedent factors and self-care, and self-care and proximal health outcomes (functional capacity and symptom burden); a descriptive, prospective component was used to examine the relationship between self-care and the distal health outcome of HRU. A convenience sample of persons with moderate or severe ACHD was recruited from a large ACHD center located in the southeastern United States. Participants completed study surveys and a six-minute walk test (6MWT) at routine clinic appointments. Clinical data were collected via medical chart review. HRU data were collected monthly for three months by self-report and validated by chart review. A total of 132 participants were recruited (mean age 36 ± 11 years, 55% female, 81% Caucasian), and 125 participants completed HRU data collection (95% retention rate).

The three manuscripts included in this dissertation research document the findings from the specific aims of this study, as well as a detailed literature synthesis on ACHD self-care. The literature synthesis documents the significant gap in current understanding of ACHD self-care, supporting the critical need for this dissertation research. The overall results of the study indicate that many persons with ACHD have suboptimal levels of self-care, and that level of self-care influences health outcomes. These results add a unique contribution to the literature both in the simple description of overall self-care behavior and in the identification of antecedents and outcomes of self-care. Further, this study adds a unique contribution in its focus on the multifaceted nature of self-care (maintenance, monitoring, and management), the use of a conceptual framework to guide self-care research (Family and Self-Care Framework), the inclusion of family influences on self-care, and the outcomes of functional capacity as measured by the 6MWT and symptom burden, which are particularly understudied in this population.

The first manuscript (Chapter 2) fills a significant gap in the literature by identifying the current state of knowledge of ACHD self-care. No known review of the ACHD self-care literature has been previously published. The review identified significant gaps in ACHD self-care knowledge, with no publications addressing the topics of overall self-care, a conceptual framework of self-care, or clinical self-care guidelines. A total of 35 publications on six individual self-care behaviors were identified. Most studies were purely descriptive and focused on physical activity or routine follow-up, with the majority of results indicating suboptimal levels of self-care. No studies reported other self-care behaviors such as medication adherence, diet and nutrition, immunization status, and the monitoring or management of symptoms. Further, special populations of ACHD, with additional self-care needs are relatively unstudied including, but not limited to, persons with heart failure, Fontan physiology, or other cyanotic groups.[3] Only 12 (34%) and 10 (29%) of studies reported antecedents or outcomes of self-care behaviors, respectively. The majority of antecedents and outcomes had an inconsistent level of evidence supporting their relationship to ACHD self-care. Only severity of ACHD and ACHD knowledge had a moderate level of evidence and no antecedents had a strong level of evidence. The only outcome with a strong level of evidence for its relationship with daily physical activity was functional capacity. No studies were identified that examined family-related antecedents or outcomes including family structure, functioning, or support, caregiver burden and caregiver health. Overall understanding of ACHD self-care is extremely limited, particularly in comparison to the similar acquired HF population in which the self-care literature is robust and self-care guidelines have already been established.[4]

The second manuscript (Chapter 3) described the findings of the first aim of the dissertation study, which was to examine the individual, clinical, knowledge, behavioral, and family antecedent factors of ACHD self-care guided by the FSC Framework. In this study, self-care (maintenance, monitoring, and management) was found to be suboptimal in the majority of participants (44.7%, 27.3%, 23.3%, respectively), supporting and extending the findings from the literature synthesis. Antecedent factors across multiple domains were significantly related to adequate level of self-care. In bivariate analysis, being older, male, having at least some college education, and having public insurance/uninsured were all moderately associated with self-care (r=.19-.37, p<.05). *Clinical factors* significantly related to self-care included NYHA Class and the Charlson Comorbidity Index. Having more comorbidities and being NYHA Class III/IV had a small to moderate association with self-care (r=.23-.30, p<.05). No statistically significant relationship was found between *knowledge* and self-care (r=.05-.06, p>.05) *Behavioral factors* significantly related to self-care included both depressive symptoms and self-care self-efficacy. Higher levels of depressive symptoms and higher levels of self-care self-efficacy had a moderate relationship with self-care (r=.24-.32, p<.05). *Family factors* significantly related to self-care included perceived family support; persons with higher confidence in family support was moderately associated with self-care (r=.18-.29, p<.05).

In multivariate analysis, factors remaining significantly related to self-care included individual (older age, higher education level, male gender), clinical (higher number of comborbidities, higher NYHA Class), behavioral (higher levels of self-care self-efficacy, more depressive symptoms), and family (higher levels of confidence in family support) factors. Factors without a significant relationship to self-care included race, insurance status, marital status, ACHD type or severity, ACHD knowledge, and parental overprotection. Further, an unreported finding in the manuscript, but an original hypothesis included under specific aim 1 of this study, was to determine if parental overprotection had a moderating effect on self-care via behavioral characteristics due to its known relationship with psychological symptoms in adulthood.[5] In addition to lacking a direct relationship to self-care, no moderating effect of parental overprotection on self-care through depressive symptoms or self-care self-efficacy was found ($\Delta R^2 = .003$, $\Delta F(1, 129) = 2.38$, $b = 0.83$, $p = .52$; $\Delta R^2 = .001$, $\Delta F(1, 129) = 2.64$, $b = -0.47$, $p = .67$, respectively).

An extensive, integrative review of antecedent factors is included in the second manuscript. Overall, the only antecedent factors with at least moderate evidence of support in previous literature, ACHD severity and ACHD knowledge, did not have a relationship with overall self-care in this study. The lack of association with ACHD severity may be related to the exclusion of persons with simple ACHD in this study in contrast to previous studies; functional severity (NYHA class and number of comorbidities) rather than defect severity may be more important for self-care abilities in persons with moderate or severe ACHD. The lack of relationship between ACHD knowledge and self-care in this study is not uncommon in other chronically ill populations, although it is recognized as an important prerequisite.[4] Overall ACHD knowledge was average in this study, slightly better than the low levels of knowledge previously reported in this population.[6-8] In multivariate analysis, adequate level of self-care was associated with nonmodifiable factors of higher education level, male gender, older age, and higher New York Heart Class, and modifiable factors of higher self-efficacy, higher depressive symptoms, and higher perceived family support for self-care. Combined with prior research, the results of this study significantly expand current understanding of antecedent factors of ACHD self-care. The majority of factors remain at an inconsistent level of understanding, primarily due to the fact that this study was the first to address those factors; two factors (ACHD severity and ACHD knowledge) changed to an inconsistent level of evidence based on the discrepant findings of this study; the lack of association between race and self-care found in this study extends prior research changing the level of evidence to moderate (Table 1). The unexpected moderate, positive association between higher depressive symptoms and better self-care monitoring should be explored further in future studies.

The third manuscript (Chapter 4) described the findings of the second aim of the dissertation study, which was to examine the relationship between self-care and health outcomes (functional capacity, symptom burden, and HRU). Mean total distance walked on the six-minute walk test was 1365 ± 275 feet. In bivariate analysis, a moderate, negative relationship was found between total distance walked and self-care monitoring (r=-.28, p=.001) and self-care management (r=-.35, p=.001). After adjusting for sociodemographic and clinical factors (age, gender, race, severity of CHD, Charlson Comorbidity Index), self-care management (β=-44.20, p=.001) and self-care monitoring (β=-14.59, p=.03) remained significantly associated with total distance walked. Persons with higher levels of self-care management and monitoring had lower functional capacity.

Participants experienced an average of 7.4 ± 4.8 symptoms in the previous month. In bivariate analysis, overall symptom burden was significantly and negatively related to self-care maintenance (r=-.29, p=.001), and positively to self-care monitoring (r=.40, p=<.001) and self-care management (r=.25, p=.02). After adjusting for clinical and sociodemographic factors, only self-care maintenance and self-care monitoring were independently associated with overall symptom burden (β=-.39, p<.001; β=1.52, p<.001, respectively). Persons with higher levels of self-care maintenance and lower levels of self-care monitoring had lower overall symptom burden.

A total of 102 (81.6%) participants experienced 310 encounters with the healthcare system. 73 (58.4%) participants experienced greater than 1 HRU encounter. In univariate analysis, self-care monitoring was significantly related to having greater than 1 HRU encounter (OR=1.21, 95% CI [1.09, 1.35], p<.001). After adjusting for sociodemographic and clinical variables, self-care monitoring remained significantly related to having greater than 1 HRU encounter and was the only variable to enter the model (OR=1.21, 95% CI [1.09, 1.35], p<.001). For every unit increase in the self-care monitoring score (indicating better self-care monitoring), the likelihood of having more than 1 HRU encounter increased by 21%. Thus, overall findings indicate that self-care was independently associated with functional capacity and symptom burden and was an independent predictor of three-month HRU.

An extensive, integrative review of health outcomes related to self-care is included in the third manuscript. Overall, this study extends previous literature by indicating a significant relationship between self-care and health outcomes. The positive relationship between self-care maintenance and symptom burden supports previous research, increasing the level of evidence to moderate (Table 1). The relationship between self-care and functional capacity and HRU, however, is inconsistent with prior studies. In this study, there was no relationship between self-care maintenance and functional capacity. Prior studies focused exclusively on physical activity levels, whereas this study focused on overall maintenance behaviors that included physical activity. Overall self-care maintenance may not be as important for functional capacity as the specific behavior of physical activity. A prior study on HRU indicated that lower level of routine follow-up was associated with increased HRU.[9] In this study, no relationship was found between overall self-care maintenance and HRU, again, most likely explained by the use of overall self-care maintenance that included routine follow-up but may not have been specific enough to detect a relationship in this study.

No prior studies examined the relationship between self-care monitoring or management and health outcomes. In this study, self-care monitoring was significantly related to all outcomes, but not as hypothesized. In all cases, worse outcomes were associated with higher levels of self-care monitoring. This finding most likely reflects that persons with higher functional limitations are more likely to perform monitoring behaviors simply because they have more symptoms that may then lead to greater, possibly protective, use of health resources. The same relationship was found between self-care management and the outcomes of symptom burden and exercise capacity, which likely has the same explanation. Future research is needed to identify protective versus harmful self-care monitoring and management.

An important, but understated contribution of this study in the manuscript was simply the description of ACHD health outcomes. Few studies have reported the symptom experience of persons with ACHD. In this study, participants experienced over seven symptoms, on average, in the prior month, and the most frequent and burdensome symptoms were identified. These findings identify a critical need to further explore and intervene to reduce symptom burden in the ACHD population. A significant amount of prior research has focused on functional capacity, but this study extends this work by its methods. The six-minute walk test, widely used in the acquired HF population, has rarely been used thus far in the ACHD population. The test is simple, inexpensive and highly correlated with cardiopulmonary exercise testing and daily activities in the acquired HF population, and may be a particularly useful, feasible test of functional capacity for both ACHD clinical and research purposes in the future. The results of this study indicate that persons with ACHD walk on average higher than persons with acquired HF, which raises questions as to the relationship between CPET and the 6MWT and its clinically significant cut-points in the ACHD population that should be explored in future research. Lastly, the findings of the HRU data show that persons with moderate or severe ACHD experience a considerable amount of HRU, even in a relatively brief follow-up period. A total of 101 (80.8%) participants experienced 310 healthcare encounters (275 office visits, 21 hospitalizations, 20 ED visits). Little is known about the preventability or protectiveness of HRU in this population, which should be explored in future studies.

Strengths and Limitations

Self-care is a critical component of chronic illness management, but the science of ACHD self-care is extremely limited. This is the first known study to address overall self-care in persons with ACHD and is thus a significant contribution to the literature. The study supports self-care as an important target for intervention by identifying suboptimal levels of self-care in the majority of participants. This exploration of antecedents and outcomes of self-care lays a foundation for building a science of ACHD self-care. Modifiable antecedent factors were identified including self-care self-efficacy and perceived confidence in family support that can be targeted for intervention development to improve self-care. Further, the identification of antecedents, both modifiable and non-modifiable, across multiple domains of influence including family factors supports the use of the Family and Self-Care Framework as a conceptual guide for building the science of ACHD self-care.

Additional strengths of this study included newly developed instruments for the ACHD population, a diverse sample, and the inclusion of a prospective component. Prior to this study, no measures of self-care or symptom burden had been developed. Both measures (Self-Care Index-ACHD [SCI-ACHD] and Congenital Heart Disease Symptom Survey [CHDSS]) had adequate face and content validity in pilot testing through expert patient and clinician consensus. In this study, internal consistency reliability for the SCI-ACHD was acceptable (self-care maintenance, CA=.62; self-care monitoring, CA=.73; self-care management, CA=.60). Reliability of the CHDSS was excellent (frequency, CA=.86; severity, CA=.87; enjoyment, CA=.89.) Although further validation is required, both the SCI-ACHD and CHDSS show promise as reliable and valid measures of ACHD self-care and symptom burden, respectively. A knowledge survey was also developed that has the potential to be an important contribution as a more feasible, content-valid measure than those previously developed. Despite the relatively small sample size, participants were almost equally distributed by ACHD complexity, gender, marital status (married/widowed versus single/living with partner/divorced), insurance type (private versus other), and education level (≤ high school degree), with diversity of age (range 21-68). The diversity of the sample increases the generalizability of findings despite the limitations of a convenience sample. Further, only 12% of eligible participants declined participation in the study, reducing selection bias. The last primary strength of this study was the inclusion of a prospective component, which allowed for the determination of a causal relationship between self-care and HRU.

Although this study has many strengths and fills a significant gap in the knowledge base of ACHD self-care, it has limitations. The primary limitations of this study are related to research design, recruitment, and measurement. First, apart from the HRU data, this study was cross sectional in nature, limiting the ability to determine causal relationships between self-care and antecedents or outcomes. The findings do confirm, however, the potential of self-care to be a modifiable factor associated with worse health outcomes, generate future hypotheses, and guide intervention development. The sample size was relatively small and a convenience sample from a single ACHD center was obtained introducing selection bias. Persons with varying levels of self-care may not have been recruited. Also, all participants were already receiving ACHD specialty care when recruited into the study. They may represent a sicker population and the findings of this study may not be generalizable to all persons with ACHD who are either not receiving any medical care or are receiving care from a non-ACHD specialist. Also, participants with recent cardiac surgery, pregnancy, physical disabilities limiting ability to walk, and adult diagnosis of ACHD were excluded from recruitment, further limiting generalizability of findings.

Regarding measurement, validated measures of ACHD self-care and symptom burden have not been previously established in the literature. Although the instruments developed by the researcher in this study were based on a well-validated instrument in the acquired HF population, they may require additional revisions to improve reliability and validity. Further, recent evidence supports a multidimensionality of the self-care maintenance and management subscales that significantly improve reliability.[57] In addition, self-care was self-reported in this study and may not reflect actual behavior. Future studies with a larger sample should investigate the dimensions of the SCI-ACHD as well as validation with objective behaviors. The ACHD knowledge survey was also developed by the researcher for this study and requires further validation in larger samples. Lastly, the sample size in this study limited the number of antecedents of self-care that could be explored. Other factors beyond the scope of this study may also be important to consider in future research including cognition, sleep, health literacy, patient-physician interactions, and general social support.[29]

Implications for Research, Practice, and Policy

Implications for Research

This dissertation study has made a significant contribution to the science of self-care, but the science remains in its infancy. Future research is needed to further establish antecedents of self-care, to confirm or disconfirm findings from this study and explore additional factors that may be important in the ACHD population. Larger cohorts and prospective studies will allow for the determination of causal relationships for more strategic and effective intervention development. Additional research is also needed to further validate the SCI-ACHD, CHDSS, and ACHD Knowledge Survey, including clinically meaningful change and cut-points for adequacy. Other health outcomes known to be related to self-care in the similar acquired HF population that were not explored in this study, including quality of life, longer-term HRU, and mortality,[4] should be explored in future studies, as well as prospective studies to further confirm the causal influence of self-care on quality and quantity of life in the ACHD population. In addition, unexpected findings in this study, such as the relationship between depressive symptoms and self-care, and symptom monitoring and health outcomes, should be further explored in future studies.

Prior to this study, no published frameworks were available to guide ACHD self-care research. The utility of the FSC Framework developed for the acquired HF population was tested in this study and may continue to be used in future research. Overall, the utility of this framework is supported by the results of this study. Antecedent factors across multiple domains (individual, clinical, knowledge, behavioral, and family) were significantly related to self-care. In particular, confidence in family support was significantly related to level of self-care, supporting the assumption that family is an essential component of self-care in chronic conditions, and in particular, conditions such as ACHD that are by nature lifelong. Future studies should continue to explore how family influences the practice and development of self-care in this population, including factors related to family structure, function, and caregiver health.

Implications for Practice

The results of this study have significant implications for clinical practice. Suboptimal self-care was prevalent in this study. Indeed, the majority of persons had inadequate levels of self-care, particularly in the area of symptom monitoring and management. Identification of self-care needs and barriers is thus an important area of assessment and intervention in the clinical setting. Although a lack of ACHD self-care guidelines makes addressing self-care more difficult, they are not necessary for individual change. Resources are available to help clinicians, including the ACHD Health Passport created by the Adult Congenital Heart Association, which outline the unique knowledge and self-care needs of patients. The Health Passport is lengthy, however, and future research should also included development of self-care tools that are feasible to use in the clinical setting. Also of note, knowledge did not adequately explain self-care in this study, and although it is acknowledged as an important prerequisite, educating patients on self-care needs is likely an insufficient intervention. This study suggests that assessing a persons' confidence in ability to meet self-care needs, identifying and treating depressive symptoms, and assessing confidence in family support of self-care needs are three important areas of assessment. The ability to translate this knowledge, however, into clinically feasible means of assessment remains a challenge to researchers and clinicians.

Implications for Policy

The results of this study have important policy implications related to self-care standardization and intervention. Currently, no evidence-based guidelines exist for ACHD self-care. A lack of guidelines may contribute to lower levels of self-care. Without guidelines, clinicians may not be addressing self-care needs or patients may be getting mixed messages about their self-care needs, which has been reported to influence physical activity levels.[10] Further, a lack of standardized self-care guidelines weakens the ability to create a strong knowledge base of ACHD self-care. Diversity in defining and measuring ACHD self-care may inhibit comparison of future studies and thus slow the science of ACHD self-care. Establishing self-care guidelines at the beginning of ACHD self-care science should be an important policy initiative. In relation to establishing guidelines, it is important to recognize and respond to the current, limited, state of knowledge of long-term outcomes in the ACHD population. As long-term outcomes become better established, self-care needs will also change. For instance, in the past 5 years, almost universal liver damage has been identified in adults with single ventricle heart defects palliated with a Fontan procedure.[11] Self-care requirements related to preserving liver health, such as tobacco and alcohol cessation and updating Hepatitis vaccinations, are now assumed to be essential parts of care for these adults. Researchers, clinicians, and policy makers must work together to quickly move new knowledge from bench to bedside to standardized care. Lastly, standardizing ACHD self-care will allow for earlier interventions to improve self-care during the difficult transition from pediatric to adult care, in which time up to 60% of young adults are lost to follow-up care.[12]

In this study, the majority of adults had suboptimal levels of self-care, yet the ability to intervene to address self-care needs is often inhibited by time and resources in the healthcare setting. This lack of time and resources is often driven by a lack of reimbursement by third parties for patient education and support in the clinical setting. Policies supporting self-care interventions are particularly important in a healthcare landscape in which ACHD care is difficult to obtain. Approximately 100 ACHD specialty clinics exist to care for an estimated 0.5-1 million adults with moderate or severe ACHD who require this level of care.[13] Patients often live far from special centers and must learn to manage and advocate for their own health, or work in collaboration with non-specialty providers. In some chronically ill populations, such as diabetes, third party reimbursement has been established for self-care education and intervention, but in most chronic conditions including ACHD, no reimbursement is available.

Future research is needed to continue to establish the relationship between self-care and health outcomes, to support policy changes in the clinical settings.

Summary

The distribution of congenital heart disease has changed dramatically over the last four decades, with the burden of disease shifting from the acute, life-threatening illness of the newborn to the chronic, life-limiting illness of the adult.[14] Evidence-based treatment and management of ACHD remains in its early stages, with the first AHA/ACC guidelines published just six years ago.[3] The role of self-care, a vital component of chronic illness management, also remains evidence-scarce in this population.

Overall, this study indicates that a large portion of persons with ACHD do not have adequate levels of self-care. Although some predictors of self-care in this study were nonmodifiable (age, gender, education level), these factors may help to identify persons at risk of low levels of self-care that may benefit most from intervention. Self-care self-efficacy shows promise as a modifiable risk factor for future interventions, as well as family support. With the increasing number of persons living with ACHD and particularly the recent growth in persons with severe ACHD, the need to develop interventions to reduce morbidities associated with long-term complications is critical. Few prior studies have examined the relationship between self-care and health outcomes. The results of this study indicate that self-care may be an important, modifiable predictor of reduced functional capacity, symptom burden, and increasing HRU in the ACHD population. This study is an important first step in expanding the evidence-base for the specific antecedents and outcomes associated with self-care in ACHD that will lead to the development of evidence-based interventions supporting self-care, and ultimately, improving both quality and quantity of life for persons living with ACHD.

Table 1. Gaps in the Literature for ACHD Self-Care

Self-Care Behaviors	Antecedents of Self-Care	Outcomes of Self-Care
Self-care Maintenance	*Individual*	*Proximal*
Routine follow-up	**Age (I)**	**Exercise Capacity (S)**
Physical Activity	**Gender (I)**	**Symptom Burden (M)**
Smoking cessation	**Race (M)**	Psychological Status
Alcohol restriction	**Marital Status (I)**	
Eat a heart-healthy diet	Employment Status	*Distal*
Endocarditis prevention:	**Insurance Status (I)**	**Quality of Life (I)**
	Education Level (I)	**Mortality (I)**
Prophylactic antibiotics	**Place of Cardiac Care (I)**	**Health Resource Use (I)-remains**
Dental hygiene		
Avoid body piercings/tattoos	*Clinical*	
	Type of ACHD (I)	*Family*
Update immunizations	**Severity of ACHD (I)**	Caregiver Burden
Maintain a healthy weight	**Body Mass Index (I)**	Caregiver Health
Medication adherence	**Blood pressure (I)**	
Pacemaker care	Ventricular Function	
Women:	**New York Heart Class (I)**	
Appropriate birth control	**Comorbidities (I)**	
Comprehensive exam prior to pregnancy	Cognitive Function	
	Knowledge/Skills	
	ACHD Knowledge (I)	
Self-care Monitoring		
Heart failure: edema checks, daily weights	*Behavioral*	
	Depression (I)	
Change in exercise tolerance	Anxiety	
	Health beliefs (I)	
Symptom recognition	**Self-efficacy (I)**	
Self-Care Management	*Family*	
Call healthcare provider	Family structure	
Take as needed medications	Family functioning	
	Family self-efficacy	
Change level of activity	**Family support (I)**	

*__BOLD__ ≥ 1 one study on topic; (I)=Inconsistent Evidence, (M)=Moderate Evidence, (S) Strong Evidence

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