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Examination of Adherence, the Quality of the Physician/Patient Relationship, and Illness-Related Beliefs Among Adults with Long QT Syndrome

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EXAMINATION OF ADHERENCE, THE QUALITY OF THE PHYSICIAN/PATIENT RELATIONSHIP, AND ILLNESS-RELATED BELIEFS AMONG ADULTS WITH LONG QT SYNDROME

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Dissertation Approval

This is to certify that the thesis presented to us by Maggie Monk on the 4th day of April, 2016, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

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Abstract

Long QT Syndrome (LQTS) is a cardiac arrhythmia disorder that affects 1 in 2,000 individuals and is a precursor to various cardiac events, including sudden cardiac arrest (Schwartz et al., 2016). As a precaution, individuals with all types of LQTS have been advised to modify their lifestyles to avoid triggers, including limiting physical activity (PA). Nonadherence to treatment recommendations could result in devastating outcomes. The purpose of the study was to explore characteristics pertaining to adherence and nonadherence to PA recommendations, including the quality of the physician/patient relationship and illness beliefs among adults with LQTS. An Internet survey was completed by 91 adults with LQTS who were recruited through various social media and medical group venues. The findings indicated that agreement on treatment goal between participants and cardiologists accounted for 3% and 4% of the variances, respectively, of perceptions of personal and treatment control over LQTS. Positive perceptions of personal and treatment control significantly predicted adherence. These results can be explained by the self-regulatory model, which describes the connection between health behaviors and illness perceptions. The findings generate many opportunities for future directions with the LQTS population and other medical populations, including research, intervention development, and advocacy.

Table of Contents

Figure

Tables

Chapter 1

Introduction

Statement of the Problem

Long QT Syndrome (LQTS) is a cardiac arrhythmia disorder that affects 1 in 2,000 individuals (Schwartz et al., 2016). It involves an irregular prolonged repolarization that results in an elongated QT interval identifiable on an electrocardiogram (Kramer & Zimebaum, 2011). The lengthened interval causes a type of dysrhythmia known as *torsades de pointes* (TdP) that makes it difficult for the heart to self-regulate and is a precursor to various cardiac events, including sudden death (Vincent, Timothy, Fox, & Zhang, 1999).

Thirteen genes have been identified that make individuals susceptible to LQTS (Vincent et al., 1999). Tthe majority of individuals with the disorder have been found to have one of three specific genes known as LQT1, LQT2, and LQT3 (Yan & Kowey, 2011). LQT1 is found in 30% to 35% of individuals with LQTS, and this type of mutation makes individuals vulnerable to experiencing a cardiac event when participating in a physical activity (PA) or when emotionally aroused (Yan & Kowey, 2011). Individuals with LQT2 most commonly experience TdP as a result of auditory stimuli, such as hearing a telephone ring or alarm clock sound (Wilde, Jongbloed, & Doevendans, 1999). Those with LQT3 experiencing TdP during periods of rest (Johnson, Hofman, & Haglund, 2009). As a precaution, individuals with all types of LQTS are advised to modify their lifestyles to avoid all triggers, including significantly limiting PA (Yan & Kowey, 2011).

In 2005, the 36th Bethesda Conference published guidelines for athletic participation for individuals who have LQTS (Zipes et al., 2005). Nonadherence to these guidelines could result in devastating outcomes for patients with LQTS, including syncope, seizures, and sudden cardiac arrest (Kramer & Zimebaum, 2011), or corrective surgeries if impact damages a device. However, studies also suggest that these guidelines may have been too stringent, and continued participation for some individuals may not result in an adverse cardiac event (Johnson & Ackerman, 2014). In addition, there is great variability in physicians' ability to diagnose, risk stratify, and treat individuals with LQTS (Taggart, Haglund, Tester, & Ackerman, 2007). Undoubtedly, additional information regarding variables associated with patients with LQTS participation in PAs is important. Information acquired through research regarding medical adherence with the LQTS, population is limited. This data may inform providers' understanding of whether patients may participate in PAs.

Adherence is typically thought of as the result of a collaborative relationship between physician and patient that results in an agreed upon treatment plan (Cramer et al., 2008). The association between a positive physician/patient relationship and greater adherence to medical recommendations has been well-documented with various medical populations, including patients with HIV (Schneider, Kaplan, Greenfield, Li, & Wilson, 2004), type I diabetes (Moine et al., 2007), prostate cancer (Orom, Hormish, Hormish, & Underwood, 2013), inflammatory bowel disease (Goldring, Taylor, Kemeny, & Anton, 2002), and hypertension (Chen, Tsai, & Chou, 2011). As a result, researchers have identified specific variables that have been shown to affect the quality of the physician/patient relationship (Schneider, Kaplan, Greenfield, Li, & Wilson,

2004). These factors include patients' perceptions of the quality of their general communication with their physician, overall satisfaction with care, and adherence dialogue (Schneider et al., 2004).

Currently, information regarding the quality of the physician/patient relationship and adherence to treatment recommendations in the LQTS population is lacking. However, a recent study found that adults with LQTS who visited the emergency room reported poor working alliance and had maladaptive problem-solving skills (Felgoise, Lawrence, & Vetter, 2013). An additional qualitative study analyzed common barriers to adherence to medical recommendations that were disclosed by individuals with LQTS and their caretakers (Steinhauser, 2010). Results demonstrated that a sense of loss of control was a common theme that was influenced by poor communication between the physician and patient, which impacted treatment adherence (Steinhauser, 2010). Specifically, participants in this study reported that they had a general feeling of having little control over the diagnostic process, treatment options and recommendations (Steinhauser, 2010).

In addition to the physician/patient relationship, research has demonstrated that illness beliefs also impact the likelihood of nonadherence to medical treatment (Moss-Morris et al., 2001). Leventhal, Diefenbach, and Leventhal (1992) developed the self-regulatory model to better understand the connection between adherence to medical treatment and illness beliefs. The model suggests that medical patients strive to maintain a state of normalcy by creating an illness identity, implementing strategies to cope with their illness beliefs, and evaluating the effectiveness of the chosen coping strategy. According to this model, illness-related behaviors, such as nonadherence to

medical recommendations, are a result of the relationship between the illness beliefs and related emotional processes. Specifically, beliefs that may impact adherence or nonadherence in medical populations have been shown to be related to illness identity, timeline of illness, illness changeability, illness consequences, treatment control, personal control, coherence, and emotional representation. Leventhal et al (1992) speculated that these illness beliefs result from experiences related to the illness, which may include interactions with physicians. Based on results of a qualitative study investigating barriers to adherence in individuals with LQTS (Steinhauser, 2010), beliefs regarding treatment control, personal control, and coherence may be impacted by the physician/patient relationship and influence levels of adherence.

Purpose of the Study

The quality of the physician/patient relationship and specific illness beliefs have been shown to predict adherence in various medical patient populations (Chen et al., 2011; Moss-Morris et al., 2001). Nonadherence to PA recommendations for individuals with LQTS could lead to serious consequences, including sudden cardiac arrest (Kramer & Zimebaum, 2011). However, information regarding nonadherence, the physician/patient relationship, and illness beliefs has not been studied with this population.

The purpose of the study was to explore the characteristics pertaining to adherence to PA recommendations, the quality of the physician/patient relationship, and illness beliefs in adults with LQTS. The quality of the physician/patient relationship was measured by overall communication (as defined by the Overall Communication Scale; Wilson & Kaplan, 2000), adherence dialogue (as defined by the Adherence Dialogue

Scale; Schneider, Kaplan, Greenfield, Li, & Wilson, 2004), patient satisfaction (as defined by the Overall Satisfaction with Care Scale; Davies & Ware 2001), and working alliance (as defined by the Working Alliance Inventory Short-Client Form; Tracey & Kokotovic, 1989). Specific illness-related beliefs assessed were patients' coherent understanding of the illness, as well as illness beliefs regarding patients' personal control of the illness and treatment control. This information is likely to be beneficial to health care professionals who work with this population and may facilitate effective patient/physician discussions concerning treatment recommendations.

Literature Review

Nonadherence to medical recommendations has been shown to have significant consequences for individuals with congenital heart disease (Yan & Kowey, 2011). The biopsychosocial model and patient-centered model suggest that factors related to specific characteristics of physicians and patients independently and the interaction between physicians and patients influences adherence (Campbell & Rohrbaugh, 2006; Epstein et al., 2005). Important factors that have been shown to be related to adherence among many medical populations includes the quality of the physician/patient relationship (Chen et al., 2011; Goldring et al., 2002; Moine et al., 2007; Orom et al., 2013; Schneider et al., 2004), illness-related beliefs (Goldring et al., 2002, Janz & Becker, 1984; Ronis, 1992), and demographic information (Turner, Laine, Cosler, & Hauck, 2003; Schneider, Kaplan, Greenfield, Li, & Wilson, 2004; Kaplan et al.,1995). Because nonadherence to recommendations regarding PA in individuals with long QT syndrome can result in cardiac arrest (Yan & Kowey, 2011), examining factors that increase the likelihood of adherence is extremely important.

Long QT Syndrome

LQTS is a life-threatening cardiac disease that is the result of irregular prolonged repolarization and a subsequent elongated QT interval (Kramer & Zimebaum, 2011). LQTS occurs in all races and ethnicities (Vincent, Timothy & Zhang, 1997), although the highest rates are among Caucasians (Schwartz et al., 2016). The prevalence of the syndrome is approximately one in 2,000 (Schwartz et al., 2016). Untreated individuals with LQTS have an increased likelihood of experiencing sudden cardiac arrest (Tester & Ackerman, 2007). Consequently, adherence to treatment is highly important for this population (Tester & Ackerman, 2007). Ongoing research has provided vast information regarding the variability of the disease, and LQTS may present differently among individuals in terms of symptom presentation, method of diagnosis, type of LQTS, and treatment recommendations (Yan & Kowey, 2011). Because symptom presentation often facilitates physicians' treatment recommendations for individuals with LQTS (Yan & Kowey, 2011), understanding the various symptoms that can be experienced is important.

Symptoms. Individuals may be diagnosed with LQTS after they experience a cardiac event (Yan & Kowey, 2011), which is most often syncope (Barsheshet et al., 2004). Additional symptoms of LQTS may include seizures, palpitations, and sudden cardiac arrest (Yan & Kowey, 2011). Typically, cardiac dysrhythmia known as *torsades de pointes* (TdP) is the precursor to a cardiac event when the heart is unable to revert back to a normal rhythm (Vincent et al., 1999). TdP is often triggered by PA or emotional arousal (Yan & Kowey, 2011). However, although information acquired through registries may be biased, ongoing observations made by the International LQTS

Registry indicated that the length of the QT interval is a strong predictor of TdP (Kramer & Zimebaum, 2011;Yan & Kowey, 2011).

Experts agree that the chances of sudden cardiac arrest double when the QT interval increases by 40 milliseconds (Kramer & Zimebaum, 2011). Additional risk factors for TdP include a history of cardiac events, having the LQT2 or LQT3 gene mutation, experiencing symptoms before 5 years of age, and being a postpubertal female (Priori, Schwartz, & Napolitano, 2003). Additionally, boys have a higher risk of sudden cardiac arrest during childhood, and females are at the highest risk during adulthood (Goldenberg et al., 2008). Research has also been conducted comparing clinical differences in African American and Caucasian individuals with LQTS (Fugate et al., 2010). Findings indicated that African American participants had a significantly longer QT interval than Caucasians, although risk of experiencing a cardiac event was similar in both groups. If a patient knows that he or she has experienced TdP or is at risk for TdP, adhering to medical treatment is important to avoid a potentially life-threatening cardiac event. Furthermore, environmental triggers for TdP are variable and specific triggers an individual with LQTS should avoid can be identified through a thorough diagnostic evaluation (Kramer & Zimebaum, 2011).

Diagnosis. Diagnosing LQTS is often challenging because the clinical and genetic presentations are highly variable (Aziz et al., 2014; Modell & Lehmann, 2005). Many individuals who are suspected of having LQTS are diagnosed as a result of genetic testing and/or clinical examination (Kramer & Zimebaum, 2011). There are numerous clinical observations that facilitate the diagnosis of LQTS, including obtaining

family history (Barsheshet et al., 2011), results of an electrocardiograph (Kramer & Zimebaum, 2011), and findings from an exercise stress test (Aziz et al., 2011).

Clinical exam. Obtaining a family history is an essential part of the clinical examination because most cases of LQTS are the result of heritable genetic mutations (Barsheshet et al., 2011). Important information obtained as part of the clinical examination includes family history of cardiac events and family history of LQTS diagnosis (Barsheshet et al., 2011). In addition to taking a thorough family history, cardiologists often recommend that patients undergo an electrocardiograph (ECG; Modell & Lehmann, 2006). An ECG records the electrical activity of the heart and allows for computer-generated measurements and interpretation (Modell & Lehmann, 2006). Individuals with LQTS may have a normal QT interval on ECG, called concealed LQTS, or they may show a borderline prolonged QT interval (Aziz et al., 2011). An ECG indicates LQTS in the general population when the QT interval is greater than 440 milliseconds (ms) in males and 460 ms in females among those who are not taking medications that could induce a prolonged QT interval (Corrado et al., 2010). Highly trained athletes may have greater values on an ECG than to the general population, and it is recommended that QT intervals exceeding 480 ms in females and 470 ms in males who are athletes be used as comparison standards (Corrado et al., 2010). All individuals who have a QT interval above 500 ms are at high risk for experiencing a cardiac event (Corrado et al., 2010). For accurate diagnosis, a scoring system is often used that includes calculating patient characteristics such as history, clinical presentation, and ECG results into a weighted sum that yields a low, intermediate, or high probability of having the condition (Modell & Lehmann, 2006; Schwartz, Moss, Vincent, & Crampton,

1993). However, with advances in genetic testing, this approach has been shown to be used less frequently (Hofman et al., 2007).

Along with ECG testing, patients who have LQTS may be advised to undergo exercise stress testing (Aziz et al., 2011; Dillenburg & Hamilton, 2002; Modell & Lehmann, 2006). Using an exercise stress test has been empirically shown to be effective in determining a LQTS diagnosis for individuals with LQT1 or LQT2, but does not determine the specific type of LQTS (Aziz et al., 2011). The exercise stress test has also been shown to be beneficial for detecting LQTS in children and adults, although caution must be used when deciding on the most effective method of diagnosing LQTS, due to the presence of a small sample size in the research (Aziz et al., 2011). In addition to a thorough clinical examination, a diagnosis is often also given as a result of genetic testing (Yan & Kowey, 2011).

***Genetic testing*.** Determining the type of LQTS an individual has is often difficult for cardiologists, due to wide variation in genotype and phenotype presentations (Kramer & Zimebaum, 2011). Genetic testing often provides important information regarding diagnosis, treatment, and risk of having a cardiac event (Barsheshet et al., 2011). For instance, research has shown that a child of a parent with LQTS has a 50% chance of having the disease, and individuals are typically encouraged to undergo genetic testing after a family member is diagnosed (Goldenberg, et al., 2008). However, although genetic testing has enhanced the ability for cardiologists to make a diagnosis of LQTS, up to one third of patients in whom a diagnosis was warranted subsequent to a clinical examination do not have the known related genes (Aziz et al., 2011; Barsheshet et al., 2011).

Information acquired through research regarding genotype-negative individuals with LQTS is limited. However, information pertaining to individuals who have known genetic mutations associated with LQTS, which is the majority of the LQTS population (Yan & Kowey, 2011), continues to grow. Thirteen genetic variations of the disease that are associated with over 600 mutations have been discovered (Goldenberg & Moss, 2008; Kramer & Zimebaum, 2011). Three types of LQTS are most prevalent and have been studied intensely (Yan & Kowey, 2011). Of all individuals with LQTS, LQT1 accounts for 30% to 35%, LQT2 is responsible for 25% to 30%, and LQT3 is present in 5% to 10% (Yan & Kowey, 2011). Genetic testing is important because 25% of individuals with LQTS types 1, 2, or 3 have a normal QT interval on an ECG test (Goldenberg et al., 2011). Undoubtedly, ongoing investigation of the clinical and genetic components of the condition is imperative to accurate diagnosis and treatment. Although further research is needed in this area, focusing on the most prevalent types of LQTS is beneficial for the majority of patients.

*LQT1***.** The majority of patients with LQTS have LQTS type 1 (Aziz et al., 2011). A normal heart adapts to faster heart rates by shortening the QT interval (Aziz et al., 2011). LQT1 results from a genetic mutation on the KCNQ1 gene, which renders the calcium channel needed for normal recovery and a shortened QT interval nonfunctional (Aziz et al., 2011). Consequently, the QT interval becomes prolonged (Aziz et al., 2011). Patients who have LQT1 typically experience cardiac events that are triggered by various PAs, including swimming and running (Albertella, Crawford, & Skinner, 2011; Modell & Lehmann, 2006) and emotional arousal, which may involve startle or fear (Schwartz, Priori, & Spazzolini, 2001).

*LQT2***.** Individuals with LQT2 have a mutation on the KCNH2 gene, which impairs a specific component of the heart needed for successful repolarization from intermediate heart rates (Aziz et al., 2011). Patients with LQT2 most commonly experience TdP and unsuccessful repolarization as a result of auditory stimuli, such as a telephone or alarm clock (Wilde et al., 1999). Fifteen percent of individuals with LQT2 also experience symptoms during periods of rest or sleep (Johnson et al., 2009).

*LQT3***.** LQT3 is the third most prevalent type and involves mutations in SCN5A genes (Chandra, Bastiaenen, Papadakis, & Sharma, 2013). At least 56 mutations on the SCN5A gene have been identified (Modell & Lehmann, 2006). Mutations associated with LQT3 differ from those related to LQT1 and LQT2 in that the first two types of LQTS result in a loss of function, while LQT3 mutations result in a gain of function associated with encoding the sodium channel (Chandra, Bastiaenen, Papadakis, & Sharma, 2013). Individuals with LQT3 typically experience a cardiac event during periods of sleep or rest, and although less likely, may also experience an event during PA (Yan & Kowey, 2011). As a result of the variability in genotype and phenotype presentations among people with LQTS, treatment protocols are also highly variable (Johnson & Ackerman, 2013; Roston, Souza, Sandor, Sanatani, & Potts, 2013)

Treatment. Due to the seriousness of symptoms that individuals with LQTS may experience, adherence to medical, surgical, and behavioral treatments is important for survival (Yan & Kowey, 2011). Cardiologists' treatment decisions are likely to depend on the type of LQTS a patient has, the patient's age, and his or her history of symptoms (Dorostkar, Eldar, Belhassen & Scheinman, 1999; ; Radico, 2013; Yan & Kowey, 2011). Medical recommendations often include pharmacological treatment (Yan &

Kowey, 2011), surgical procedures (Dorostkar et al., 1999), and lifestyle modifications (Fitzgerald & Ackerman, 2005).

Medications. Pharmacological treatment of LQTS often involves avoiding specific medications, as well as being prescribed new ones (Yan & Kowey, 2011). Patients are typically encouraged to discontinue prescriptions that may elongate the QT interval, including specific anti-arrhythmics, antibiotics, motility agents, narcotics, decongestants, and psychotropics (Yan & Kowey, 2011; Bennett, 2013). Patients who do not experience LQTS-related symptoms, but are on medications that can precipitate an LQTS-related event (e.g. Albuterol for asthma), are confronted with a difficult decision regarding discontinuing their current prescriptions. Although discontinuing medications for conditions other than LQTS may result in significant negative consequences, some experts have argued that the benefits of adhering to LQTS pharmacological treatment outweigh these costs (Yan & Kowey, 2011).

Along with discontinuing medications that result in an elongated QT interval, taking medications that enhance the repolarization process is often recommended (Kramer & Zimetbaum, 2011). Medications that are often prescribed to patients with LQTS include beta blockers, potassium supplements, sodium channel blockers, and calcium channel blockers (Kramer & Zimebaum, 2011). Adherence to these medications has been shown to reduce the risk of a sudden cardiac arrest by 75% in LQT1 and 50% in LQT2 (Goldenberg et al., 2008). If a patient requires additional treatment in conjunction with medication, surgical procedures are typically considered (Yan & Kowey, 2011).

Surgical procedures. Invasive therapies are often used in patients who have greater risk of sudden cardiac arrest (Albertella et al., 2011). Some individuals are given

implantable cardioverter defibrillators (ICD) when they are unable to take beta blockers (Albertella et al., 2011; Moss et al., 2000) or when they have experienced sudden cardiac arrest (Yan & Kowey, 2011). A recent study found that there was a 1.3% death rate among patients with LQTS who were considered to be high-risk for having cardiac arrest and had ICDs, compared to a 16% death rate in such patients who did not have ICDs (Zareba et al., 2003). Consequently, use of an ICD is considered a primary prevention technique (Cesario & Dec, 2006).

There are various surgical alternatives to using an ICD (Yan & Kowey, 2011). Physicians may recommend the implantation of a pacemaker to prevent TdP, and research supports using a combination of beta blocker therapy, a pacemaker, and ICD for younger patients who tend to be less medically adherent (Dorostkar et al., 1999). If a patient decides against having a pacemaker and/or ICD, the implications could be fatal (Dorostkar et al., 1999). Along with pharmacological and surgical treatment for LQTS, behavioral modifications are also recommended to avoid future cardiac events (Fitzgerald & Ackerman, 2005).

***Lifestyle modifications*.** Persons with LQTS are typically encouraged to make lifestyle changes to decrease the chances of TdP and subsequent cardiac arrest (Fitzgerald & Ackerman, 2005). The changes often include engaging in healthy dietary habits, such as consuming potassium enriched foods and staying hydrated (Fitzgerald & Ackerman, 2005). Patients are also asked to avoid substances that increase the QT interval, such as purple grapefruit juice (Fitzgerald & Ackerman, 2005), the supplement cesium chloride, amphetamines (Vyas, Johnson, Houlihan, Bauer, & Ackerman 2006), and caffeine (Rottlaender, Motloch, Reda, Larbig, & Hoppe, 2012). Along with making dietary

changes, LQTS patients may be told to avoid triggers that could result in TdP, such as startling auditory stimuli and high impact PA (Zipes et al., 2005). These may be difficult adjustments for a patient whose job requires being in a loud and strenuous environment. In addition, physicians recommend that an identification card be worn at all times by patients with LQTS and that families take an automatic external defibrillator on all outings (Yan & Kowey, 2011). Furthermore, due to the risk of sudden cardiac arrest after PAs in persons with certain types of LQTS, cardiologists often recommend changes in PA participation (Yan & Kowey, 2011).

PA restrictions. The 36th Bethesda Conference published the accepted PA restrictions for individuals with arrhythmias, including LQTS (Zipes et al., 2005). These guidelines are largely based on data obtained from nonathletes, the general perceptions of the medical community (Zipes et al., 2005), and expert consensus (Roston et al., 2013). The guidelines recommend that all individuals with electrocardiographically confirmed LQTS avoid participation in most competitive sports (Zipes et al., 2005). Individuals who are diagnosed with LQT3 are recommended to avoid all competitive sports, while individuals who are diagnosed with LQTS by genetic testing and are asymptomatic with normal QT intervals are permitted to participate in competitive sports (Zipes et al., 2005). This set of guidelines also discourages participation in sports with a danger of bodily collision for individuals with an ICD (Zipes et al., 2005). In addition to competitive sports, there are also limitations for various PAs of high, moderate, and low intensities.

Of the numerous activities that increase the risk of a cardiac event in individuals with LQTS, swimming has been shown to be highly associated with having an event

(Modell & Lehmann, 2006). A Mayo Clinic study demonstrated that of 388 individuals with LQTS, 28 reported a history of cardiac events while swimming (Choi, Porter, & Ackerman, 2004). Of these, 85% had LQT1 (Choi et al., 2004). Hypotheses addressing the exact mechanisms that result in cardiac arrest due to swimming are controversial (Choi et al., 2004; Goldenberg et al., 2008; Gyorke, 2009; Katagiri-Kawade, 1995). However, neglecting the increased prevalence of sudden death related to being in water for individuals with LQT1 could have devastating consequences.

Consequences of nonadherence. Nonadherence to PA recommendations could result in tragic outcomes for patients with LQTS, including syncope, seizures, and sudden cardiac arrest (Kramer & Zimebaum, 2011). The United States registry data from 2009 indicates that there is a 2% incidence of sudden cardiac arrest in athletes due to LQTS (Maron, Doerer, Hass, Tierney, & Mueller, 2009). However, expert opinions regarding PA treatment recommendations have been mixed. The risk of having a cardiac event when engaging in PAs for individuals with LQTS has been well documented (Choi, Porter, & Ackerman, 2004; Modell & Lehmann, 2006). An activity that increases the QT interval to 550 ms is thought to be a high-risk activity for individuals with LQTS (Modell & Lehmann, 2006). However, the medical community typically encourages regular PA to avoid heart disease, diabetes, and other serious medical conditions (Shipman, 2010). Additionally, a recent study that used records of patients between the ages 6 and 40 who were previously evaluated at the Mayo Clinic's LQTS Clinic from 2000 to 2010 investigated the outcomes of those who chose to continue participating in competitive running (Johnson & Ackerman, 2014). Participants were 130 individuals, including 20 participants with ICDs (Johnson & Ackerman, 2014). Of the 130 participants, 70 had an

LQTS-related genetic mutation without the presence of an elongated QT shown on an ECG or history of cardiac events, and of these, none had a sports-related cardiac event during the study (Johnson & Ackerman, 2014). The remaining 60 athletes also had an LQTS-related genetic mutation, but had an elongated QT interval on an ECG and history of cardiac events. Of these, one had a sports-related cardiac event and was revived by an ICD shock (Johnson & Ackerman, 2014). Overall, the findings suggested that athletes are capable of making appropriate decisions regarding their participation in competitive sports after LQTS diagnosis (Johnson & Ackerman, 2014). This is supported by previous research that suggested that when medical patients manage their own illness, it increases positive health outcomes (Michie, Miles, & Weinman, 2002). Furthermore, these results suggest that the 36th Bethesda Conference guidelines regarding PA for individuals with LQTS may be too stringent (Johnson & Ackerman, 2014).

An additional study found that Canadian physicians, who are required to adhere to similar guidelines to American physicians only partially implemented the current recommendations (Roston et al., 2013). These results indicate that there is widespread disagreement regarding the applicability of the current guidelines. Undoubtedly, recommending the appropriate treatment regimen for individuals with LQTS is challenging for physicians.

Physician decision-making. A recent qualitative study evaluated themes regarding physician decision-making when treating individuals with LQTS (Radico, 2013). Five themes were identified; continuous reevaluation of decisions, impact of physicians' experience with treating patients with LQTS, being different from or joining other physicians, special considerations regarding patients' involvement in PAs, and

physicians as recommenders rather than decision-makers (Radico, 2013). Additionally, physicians conveyed that their decisions regarding treatment were often based on the patient's age, particularly when treating children or adolescents, who may have difficulty accepting the need to decrease participation in PAs (Radico, 2013). Because recommendations regarding participation in PAs may require patients to make difficult behavioral changes, understanding factors that are likely to result in nonadherence may contribute to important information for healthcare providers who work with this population.

*Adherence barriers***.** Barriers to medical treatment adherence have been identified among various medical populations. Understanding how cultural variables enhance or impede the physician/patient relationship is important when examining adherence to medical recommendations (Engle, 2007; Epstein et al., 2005). Studies of various medical populations have found that cultural characteristics, such as race, age, gender, and socioeconomic status have been shown to impact medical adherence (Cooper et al., 2003; Kaplan et al., 1995; Schneider et al., 2004; Turner et al., 2003). Continuing to investigate cultural differences among adults with LQTS is be important to include in future studies.

Although cultural considerations and adherence have not been studied in adults who have LQTS, a recent qualitative study investigated other themes related to adherence in this population (Steinhauser, 2010). In regard to the diagnostic process, many patients reported that they did not know what type of LQTS they had and therefore did not know what specific triggers represented the greatest risk for having a cardiac event (Steinhauser, 2010). Furthermore, many of the participants did not understand the

information communicated by their physicians and reported that it was often unclear and vague (Steinhauser, 2010). For example, one woman with LQTS stated that her physician told her that she could continue participating in competitive running to maintain her general health as long as she did not push herself (Steinhauser, 2010). Other participants stated that they were confused about the disease severity and did not believe that their physicians were knowledgeable enough about LQTS to effectively treat the condition (Steinhauser, 2010). This perceived uncertainty and vagueness of information communicated by their cardiologists was not only likely to increase patients' risk for engaging in PAs that could lead to a cardiac event, but also resulted in significant levels of anxiety and difficulties in decision-making, coping, and interpersonal interactions with family members (Steinhauser, 2010).

Additionally, three specific types of adherence emerged in the study that were classified as precompliance, hypercompliance, and pseudocompliance (Steinhauser, 2010). Precompliance was described as engaging in behaviors that are consistent with treatment recommendations before a formal diagnosis of LQTS was given. Behaviors that were implemented beyond what was recommended by physicians were classified as hypercompliant. Pseudocompliance was characterized as behaviors that were thought by the individual to be compliant, but actually irrelevant to treatment. Steinhauser (2010) also found that loss of control, perceptions that the disease was not manageable, and the perceived severity of the disease were recurrent themes that were related to non-adherent behaviors. Understanding how factors associated with nonadherence are related to the currently accepted medical models of patient care could guide future research in medical settings.

Models of Patient Care

Biopsychosocial model. George Engel published an article discussing the need for a new medical model (Engel, 1992). Engel's proposed model was based on general systems theory, which suggests that each system impacts and is affected by other systems (Engel, 2007). The systems that addressed were the biological, psychological, and social systems (Engel, 1992), in contrast to the traditional biomedical model centered mainly on the biological aspects of illness. The biological functions in the biopsychosocial model emphasize the anatomical, structural, and molecular basis of disease and the impact on patients' biological functioning (Campbell & Rohrbaugh, 2006). The psychological aspects of the model address how developmental factors, motivation, and personality impact patients' experiences and reactions to medical conditions (Campbell & Rohrbaugh, 2006). Additionally, the social factors included the cultural, familial, and environmental influences on patients' experiences of illness (Campbell & Rohrbaugh, 2006). Engle (1992) suggested that obtaining information relevant to all three systems facilitates healthcare professionals' understanding of patients' presenting symptoms by conceptualizing the information in a meaningful way, while also guiding the development of a comprehensive care management plan for treatment (Campbell & Rohrbaugh, 2006). Addressing nonadherence to medical recommendations using the biopsychosocial model is likely to facilitate gaining a true understanding of a patient's decision to adhere or not adhere to treatment and guide useful health-promoting interventions. The biopsychosocial model is often incorporated in physician, nurse, social work, and psychology training programs (Campbell & Rohrbaugh, 2006). Additionally, this model facilitated the development of the patient-centered model (Mead & Bower, 2000).

Patient-centered model. Patient-centered care typically refers to actions toward patient-centeredness, such as interpersonal behaviors, medical interventions, and innovations in health systems (Epstein et al., 2005). When addressing patient-centered care factors, patient characteristics, physician characteristics, clinician characteristics, and health system characteristics must all be considered (Epstein et al., 2005). The need for physicians to have a patient-centered communication style is an important component of the patient-centered model (Mead & Bower, 2000). Patient-centered communication typically refers to consideration of patients' needs, perspectives, and experiences, while providing patients with opportunities to offer input into their care (Epstein et al., 2005; Mead & Bower, 2000). Specific elements of patient-centered communication include duration of the visit, duration of patient and physician speech, and the socioemotional and psychosocial characteristics of the dialogue (i.e., rapport building, providing psychosocial information, social talk, and use of open-ended questions; Cooper et al., 2003).

Additionally, patient-centered communication is thought to be both a trait (i.e., overall communication style) and a state (i.e., behaviors during a specific interaction; McWhinney, 1995; Roter et al., 1997), and various factors have been shown to be related to the quality of patient-centered communication. Patient characteristics that have been found to impact physicians' behaviors include illness severity, personality, medical history, culture, socioeconomic status, and mood (Epstein et al., 2005). Physician factors that influence patient-centered communication include personality, risk aversion, support of patient autonomy, knowledge of patient characteristics, and having a patient-centered orientation (Epstein et al., 2005). Factors that have been found to be related to the interaction of physician and patient characteristics include race concordance, duration of

the relationship, trust, and concordance of beliefs and values (Epstein et al., 2005). Measuring patient-centered care with all factors is challenging because it involves obtaining several points of view (Epstein et al., 2005). Additionally, patient and physician self-reports are not typically highly correlated with objective ratings of the same encounters (Epstein et al., 2005).

The biopsychosocial model and patient-centered model both lend themselves to addressing adherence to medical treatment recommendations by using a multifaceted approach. Research that is guided by these models should incorporate measures of patient characteristics, such as demographic information, illness beliefs, and patient ratings of physician characteristics that are likely to impact the physician/patient relationship. It is also important for investigators to clearly define adherence in a way that portrays a patient-centered approach.

Medical Adherence and Compliance

Medical nonadherence/non-compliance is a significant problem in the United States, resulting in $100 billion in annual costs (WHO, 2003). Studies have been conducted with various medical populations to better understand factors that contribute to this problem (Cutrona et al., 2010). Adherence and compliance have been operationalized in various ways in the research, and investigators have not agreed on one specific description or method of measurement (Sankar, Nevedal, Neufeld, & Luborsky, 2007). However, adherence is usually defined as the degree to which provider recommendations about medication use are followed by the patient (Cramer et al., 2008). Compliance is thought to suggest that a patient is a passive participant in his or her treatment decisions and must follow the physician's recommendations without

question (Cramer et al., 2008). Nonadherence and noncompliance are considered to be the opposite of these definitions (Cramer et al., 2008). For the purpose of the present study, the terms adherence and nonadherence will be used to foster a patient-centered approach to addressing health behaviors.

In addition to developing various definitions of adherence, researchers have also differentiated the different types of adherence. Meichenbaum and Turk (1987) described three categories of adherence, including prescription and drug taking, health behavior changes, and treatment attendance and participation. They also noted that adherence is a complex issue that changes over time. The Patient-Centered Model encourages addressing medical nonadherence by obtaining patient and provider information (Epstein et al., 2005). Current research has found that specific patient characteristics (Goldring, Taylor, Kemeny, & Anton, 2002), physician characteristics (Cutrona et al., 2010; Radico, 2013), and factors related to the physician/patient relationship (Sabate, 2003) are all highly associated with medical adherence.

Patient characteristics related to adherence. Assessing patient characteristics is important when conceptualizing nonadherence to medical treatment with the patient-centered model and biopsychosocial model (Epstein et al., 2005; Goldring et al., 2002). There are many patient factors that are likely to impact adherence, and it is impossible to evaluate all possible factors simultaneously. Therefore, researchers must choose the factors that are likely to be the most relevant to a specific population. Recent qualitative studies have indicated that illness-related beliefs and symptom experience are likely to impact adherence rates among individuals with LQTS (Steinhauser, 2010).

***Illness beliefs*.** Beliefs that medical patients have about their conditions have been shown to influence adherence levels (Goldring et al., 2002). Numerous studies have indicated that health-related beliefs explain up to 40% of the variance in health protective and maintenance behaviors including adherence (Goldring et al., 2002; Janz & Becker, 1984; Ronis, 1992), which has led to the development of different health belief models (Goldring et al., 2002).

Leventhal and coworkers developed the self-regulatory model (SRM) to explain how illness beliefs impact behavior (Leventhal et al.,1992). Leventhal and colleagues (1992) used the SRM to better understand the connection between adherence to medical treatment, the experience of symptoms, and illness perceptions. The model suggests that individuals strive to maintain a state of normalcy by creating an illness identity, implementing strategies to cope with related cognitions, and evaluating the effectiveness of the chosen coping strategy. This process involves the development of a symptom-based schema (Leventhal et al., 1992). A schema is an organized, biased pattern of thoughts that develop as a response to experiences acquired over time (Beck, 1995). According to the SRM, illness-related behaviors are a result of the relationship between the schema and emotional processes (Leventhal, Diefenbach, & Leventhal, 1992). Qualitative research has demonstrated that illness beliefs impact nonadherence in patients with LQTS (Steinhauser, 2010), providing support for using Leventhal's SRM when conducting research assessing adherence in this population.

Illness-Perception Questionnaire-Revised. The SRM has been formulated into the Illness-Perception Questionnaire-Revised (IPQ-R; Moss-Morris et al, 2002), which is a questionnaire that is divided into sections. The first section comprises a symptom

checklist that measures illness identity and includes symptoms related to pain, nausea, fatigue, headaches, and upset stomach. The authors of the IPQ-R encourage researchers to modify this section to reflect the symptoms of the medical population of interest (Moss-Morris et al., 2002). The second section of the IPQ-R includes 50 items that address the following: timeline of illness, illness changeability, illness consequences, treatment control, personal control, illness coherence, and emotional representation. Validity and reliability of the IPQ-R have been demonstrated through extensive research with patients from various medical populations, including individuals with asthma, diabetes, rheumatoid arthritis, chronic pain, acute pain, myocardial infarction, multiple sclerosis, and HIV (Moss-Morris et al., 2002). Specifically, the IPQ-R has demonstrated high structural validity, internal reliability, test-retest reliability, discriminant validity, predictive validity, and interrater reliability (Moss-Morris et al., 2002). Furthermore, although the IPQ-R has not been used with individuals with LQTS, it has been shown to be a useful instrument when assessing illness-related beliefs among other cardiac populations (Cooper, Lloyd, Weinman, & Jackson, 1999; Weinman, Petrie, Sharpe, & Walker, 2000). .

The IPQ-R and cardiac populations. Similar to the LQTS population, medical treatment for the general cardiac population often includes pharmacological treatment, as well as recommendations for behavioral change to decrease the likelihood of sudden cardiac arrest (Carney et al., 2010). One study used this instrument to assess the connection between illness perceptions and adherence, while also taking into account the occurrence of cardiac symptoms (Cooper et al.,1999). Results suggested that there was not a relationship between illness beliefs and symptom presentation (Cooper et al.,

1999). The investigators speculated that the reason for this result may be that their data were biased by history (Cooper et al.,1999). However, belief that the medical condition was controllable was shown to predict adherence to a cardiac rehabilitation program, while the participants' belief that heart condition was related to their lifestyle predicted nonadherence to the treatment regimen (Cooper et al.,1999). Although this study provides interesting information regarding illness perceptions and adherence, the specificity of the population studied (i.e., patients with myocardial infarction who were referred for rehabilitation) may limit generalizability to other cardiac populations, such as LQTS.

In addition, a recent cross-sectional study evaluated the relationships between illness perceptions as measured by the IPQ-R and adherence to medication recommendations among 355 patients with hypertension (Chen et al., 2011). The results indicated that beliefs concerning control of the illness and beliefs concerning the cause of the illness were likely to impact patient adherence to medical recommendations (Chen et al., 2011). The authors suggest that the results support the use of the common-sense model (Leventhal, Brissette, & Leventhal, 2003), a precursor to the SRM, in explaining the role of illness identity in adherence (Chen, Tsai, & Chou, 2011).

An additional study that focused on patients with hypertension demonstrated that those with lower emotional representation and less perceived personal control were more likely to adhere to medical recommendations (Ross, Walker, & MacLeod, 2004). In another study, significant predicting factors related to adherence to medications for hypertension included beliefs regarding treatment control, risk factors, psychological attribution, symptoms experienced after diagnosis, and beliefs regarding personal control

(Chen, Tsai, & Lee, 2009). Participants in this study were from Taiwan, and the results may not generalize to other populations. However, this study provides evidence suggesting illness-related beliefs impact adherence in patients of various cultures. Along with illness perceptions, symptom experience has also been shown to influence adherence in medical populations (Goldring et al., 2002).

Symptom experience. There is a range of clinical presentations in the LQTS population, and understanding symptom history may be important for developing a sound theory as to why a particular patient chooses whether to adhere to medical recommendations (Leventhal et al.,1992). Studies have demonstrated that physical symptoms of a medical condition influence adherence to treatment (Goldring et al., 2002). Although quantitative research investigating factors related to adherence has not been conducted with the LQTS population, other cardiac populations have been studied (Carney et al., 2010). One such study demonstrated that adherence to medical treatment was greater in individuals who experienced symptoms related to their illness (Haynes & Sackett, 1979). Conversely, a more recent study investigating the impact of symptoms on adherence in patients with coronary heart disease demonstrated that patients who experienced symptoms were less adherent to medical treatment than those who did not experience symptoms (Carney et al., 2010). The authors suggested that adherence may predict symptom occurrence, rather than symptoms as a predictor of adherence (Carney et al., 2010). Because the findings in the literature are inconsistent, it is likely that symptom experience is not the only predicting factor related to adherence to medical treatment in cardiac populations. Therefore, along with obtaining patient information when assessing factors related to nonadherence, including illness-related beliefs and

symptom experience, physician characteristics should also be examined (Curtrona et al., 2010).

Physician characteristics related to adherence. An important characteristic of physicians that has been found to be related to patient adherence and nonadherence is physician participation in the administration of interventions (Cutrona et al, 2010; Goldring et al., 2002). However, the current literature addressing these variables is inconclusive. A recent meta-analysis of 82 studies that evaluated medication adherence interventions for cardiovascular disease and diabetes assessed the effectiveness of interventions with varying physician roles (Cutrona et al., 2010). The results indicated that physicians who were not involved in an intervention had a medium to large effect on adherence compared to studies that included physician involvement (Cutrona et al., 2010). Interventions considered to have no physician involvement included educational interventions, reminders of adherence, behavioral interventions guided by other professionals, and a combination of these interventions (Cutrona et al., 2010). Among the studies that required physician participation, those that incorporated passive interventions were most effective (Cutrona et al., 2010). Overall, the findings suggest that existing physician-based interventions have been less effective than methods using other healthcare professionals (Cutrona et al., 2010). Limitations of the meta-analysis include the use of published randomized controlled trials, a heterogeneous sample, and low generalizability to non-cardiovascular patients.

Conversely, research has also shown that the firmness of physicians' suggestions to take medications for patients with inflammatory bowel disease predicted higher levels of intent to adhere to medication recommendations (Goldring et al., 2002). This

discrepancy emphasizes the need for further evaluation of the current methods used to increase the likelihood of medical adherence, in addition to implementation of new methods that involve other healthcare professionals. Additionally, these studies did not address the role of other variables that are likely to impact adherence, such as the role of the physician/patient relationship (Cutrona et al, 2010; Goldring, Taylor, Kemeny, & Anton, 2002), which may have provided a possible explanation of the results.

Patient/physician interaction characteristics related to adherence. Along with characteristics specific to the patient and physician respectively, factors related to the interaction between the physician and patient are also important to identify when evaluating factors that impact adherence among individuals with LQTS (Sabate, 2003). An important component of the physician/patient interaction is the quality of the physician/patient relationship. Quality of the physician/patient relationship has been defined in varying ways throughout the literature (Sabate, 2003; Epstein et al., 2005).

Quality of the physician/patient relationship. Research has shown that this construct is likely to be impacted by specific physician variables, characteristics of the patient, and characteristics of the interaction between physician and patient (Sabate, 2003). Variables that have been incorporated into numerous studies evaluating the physician/patient relationship are general communication (Cooper et al., 2003; Kaplan, et al., 1995), satisfaction (Mead & Bower, 2000; Krupat et al., 2001; Hjortdahl & Laerum, 1992) , adherence dialogue (Schneider et al., 2004), and demographic variables (Cooper et al., 2003; Cooper-Patrick et al., 1999; Kaplan et al., 1995; Schneider et al., 2004).

General communication. Communication and a strong relationship between the physician and patient have been linked to improved quality of care, positive health

outcomes, and adherence (Shipman, 2010). However, a strong relationship can be defined in multiple ways. For instance, when patients perceive an open line of communication with their physicians, they are likely more likely to discuss important issues related to their medical condition and receive the best care possible (Shipman, 2010). These patients are also likely to have a greater understanding of their illness and related treatment options, resulting in making health-promoting decisions (Shipman, 2010). A meta-analysis of studies evaluating the impact of physician communication and patient adherence found a 19% increased risk of nonadherence among patients who reported that their physicians communicate poorly (Haskard, Zolnierek & Dimatteo, 2009). As previously stated, the patient-centered model involves multiple factors related to physicians' communication with patients (Cooper et al., 2003). Two essential components of quality communication that are in line with this model include participatory decision-making style of the physician and adherence dialogue. Participatory decision-making is defined as the tendency of physicians to make treatment decisions collaboratively with patients by providing a sense of control over treatment decisions, involving patients in treatment decisions with an adherence dialogue, and encouraging patients to have a sense of responsibility over health behaviors (Cooper et al., 2003; Kaplan, Gandek, Greenfield, Rogers, & Ware, 1995). This style of communication has been shown to produce better health outcomes than in patients of physicians who do not use collaborative dialogue (Kaplan et al., 1995). Additionally, patients who had seen their physicians for longer periods reported a greater participatory style of their physician (Kaplan, et al., 1995). Along with general communication, patient satisfaction is also commonly measured in research when evaluating the quality of

the physician/patient relationship (Spernak, Moore, & Hamm, 2007).

Patient satisfaction. The use of patient satisfaction ratings to measure patient-centered care is controversial. Researchers have suggested that the use of patient satisfaction ratings is problematic because patients who are less participatory due to culturally related beliefs regarding physicians' role are also less attuned to physicians' deficiencies (Krupat et al., 2001; Mead & Bower, 2000). Research has also shown that satisfaction does not necessarily reflect patients' trust and recommendation of their physicians (Krupat et al., 2001). Furthermore, researchers have speculated that improved satisfaction does not relate to health outcomes (Michie et al., 2003), and patients may rate higher levels of satisfaction with physicians they have seen for a longer period of time as a result of familiarity effects (Michie et al., 2003). Conversely, research has also demonstrated that continuity of medical care increases patient satisfaction and promotes positive health outcomes (Hjortdahl & Laerum, 1992; Spernak, Moore, & Hamm, 2007).

Despite this controversy, patient satisfaction is a common variable that is used when assessing adherence to medical treatment. A recent study evaluated the relationship between depression and adherence to postoperative cardiac rehabilitation treatment among 92 patients (Spernak et al., 2007). Findings demonstrated that the relationship between depression and adherence was mediated by lower satisfaction with physician interactions (Spernak et al., 2007). The authors suggested that adherence among patients undergoing cardiac rehabilitation may improve by enhancing the quality of the physician/patient interaction (Spernak et al., 2007). Along with patient

satisfaction, a strong working alliance between the patient and physician has been shown to impact medical adherence (Fuertes et al., 2007).

Working alliance. Working alliance is defined as the extent to which there is patient-provider agreement on specific goals for treatment (Bordin, 1979; Fuertes et al., 2007). This involves collaboration on explicit tasks that are put in place to work toward treatment goals (Bordin, 1979; Fuertes et al., 2007). Additionally, a working alliance involves the emotional bond between patient and provider, which is characterized by a mutual liking and trust (Bordin, 1979; Fuertes et al., 2007). The working alliance construct has been examined in both psychiatric and medical populations (Fuertes et al., 2007; Greenson, 1967) and has been considered an essential component of the patient-centered care model (Mead & Bower, 2000). Researchers believe that when patients perceive a strong working alliance with their physicians, they are more likely to see the value of treatment and feel more capable of making behavior changes (Fuertes et al., 2007).

The working alliance construct has been measured by using the Working Alliance Inventory (WAI) across theoretical approaches and with various populations (Wampold, 2000) and has demonstrated high interrater reliability, internal consistency, predictive validity, and concurrent validity (Hatcher & Gillaspy, 2005; Horvath & Greenberg, 1989). Tracey & Kokotovic, 1989; Findings have consistently shown that the working alliance is the most reliable predictor of outcome across studies (Gelso & Carter, 1985; Wampold, 2000). Additionally, the WAI has been modified in medical settings and shown to be an effect measurement tool when assessing the patient/physician alliance in cardiac populations (Evon & Burns, 2004) and has also been demonstrated to correlate

with patient satisfaction and adherence to medical treatment (Fuertes et al., 2007). Findings from a recent study that evaluated adults with LQTS who visited the emergency room indicated that the sample reported a poor working alliance with their physicians (Felgoise, Lawrence, & Vetter, 2013). Further research is needed in this area to determine the factors contributing to a poor working alliance and consequences. Along with measuring patient characteristics, physician characteristics, and the physician/patient relationship, demographic variables of the patient and physician could also impact the quality of their interactions (Kaplan et al.,1995).

Cultural variables. There are various hypotheses addressing how cultural variables influence the physician/patient relationship and adherence to medical treatment. Studies have suggested that patient characteristics such as gender, age, and race impact adherence (Kaplan et al., 1995; Schneider et al., 2004; Turner et al., 2003). One possible explanation for these results is that these demographic characteristics impact physicians' interpersonal behaviors, resulting in increased or decreased collaborative behaviors during office visits (Kaplan et al.,1995). This possibility is important to consider with medical populations, given the increased chance of positive health outcomes in patients who have collaborative dialogues with their physicians (Kaplan et al.,1995; Ong, De Haes, Hoos, & Lammes, 1995).

Along with these factors, race concordance of the physician and patient has also been shown to be associated with the quality of the physician/patient relationship (Cooper et al., 2003). Studies initially demonstrated that African American patients who saw physicians of the same race viewed medical visits as more satisfying and participatory than participants who saw physicians of other races (Kaplan et al.,1995). Additional

research demonstrated that race-discordant medical visits often result in patient reports of less involvement in decision making, lower levels of satisfaction with care, and lower levels of trust in physicians (Cooper-Patrick et al., 1999). To expand upon these results, researchers compared patient/physician communication in race-concordant and race-discordant visits by focusing on patient satisfaction and participatory decision making style of physicians (Cooper et al., 2003). The findings indicated that race-concordant visits were longer (Cooper et al., 2003). Additionally, the results demonstrated that the relationship between race-concordance and higher patient ratings of care was independent of patient-centered communication (Cooper et al., 2003). The authors speculated that patient and physician attitudes may have mediated this relationship (Cooper et al., 2003). Undoubtedly, associations between race-concordance and the physician/patient relationship may be significant. However, it is important to note that other variables, such as geographic location, socioeconomic status, characteristics of different medical facilities, and gender concordance may also mediate this relationship (Cooper-Patrick et al., 1999). Therefore, understanding how cultural variables influence the physician/patient relationship, along with other additional patient and physician factors, is important when examining adherence to medical recommendations in various populations, including adults with LQTS.

Quality of the physician/patient relationship and adherence. A combination of general communication, patient satisfaction, and adherence dialogue measures has been used to assess the physician/patient relationship, along with other measures (Schneider et al., 2004). A study evaluated these specific factors of the physician/patient relationship, along with trust in physician and willingness to recommend the physician

(Schneider et al., 2004). The study assessed the relationship between the quality of the physician/patient relationship and adherence to antiretroviral therapy for 552 individuals with HIV infection, while also assessing the impact of beliefs about antiretroviral therapy (Schneider et al., 2004). Additional information obtained from participants included gender, race, education level, age, sexual orientation, and health insurance (Schneider et al., 2004).

Results demonstrated that all measures evaluating the physician/patient relationship were significantly associated with medication adherence, with Spearman correlation coefficients ranging from 0.13 to 0.21 (Schneider et al., 2004). This supports the use of the specific satisfaction, communication, and adherence dialogue scales that were used when assessing the quality of the physician/patient relationship in future research. Additionally, participants who believed that antiretroviral therapy was important had better adherence than those who did not (Schneider et al., 2004). A weakness of the study was the use of self-reported adherence and thus a possible overestimation of adherence. However, this is a limitation of any self-report measure of adherence, and measuring adherence without using participant self-report is difficult. Overall, this study demonstrates the importance of considering general communication, satisfaction with physician, adherence dialogue, health-related beliefs, and demographic variables when evaluating adherence in medical populations.

Conclusion

Adherence has been studied in multiple medical populations over the years (Chen et al., 2011; Goldring et al., 2002; Moine et al., 2007; Orom et al., 2013; Schneider et al., 2004) As a result, researchers have obtained information that has guided the development

of various definitions of adherence by applying the information to different medical models, including the biopsychosocial model and patient-centered model (Campbell & Rohrbaugh, 2006; Epstein et al., 2005). According to these models, factors related to physician and patient characteristics independently and the interaction between physicians and patients influence adherence (Campbell & Rohrbaugh, 2006; Epstein et al., 2005). Specifically, the quality of the physician/patient relationship (Chen et al., 2011; Goldring et al., 2002; Moine et al., 2007; Orom et al., 2013; Schneider et al., 2004), illness-related beliefs (Goldring et al., 2002; Janz & Becker, 1984; Ronis, 1992), and cultural factors have been shown to be highly associated with adherence (Kaplan et al.,1995; Schneider et al., 2004; Turner et al., 2003). These specific variables are likely to influence adherence rates in various medical populations, including adults with LQTS. Information obtained from LQTS adherence studies could potentially be useful for medical professionals who work with this population by elucidating possible factors that can increase adherence rates and reduce the risk of a cardiac event.

Chapter 2

Research Questions

What characteristics are present regarding adherence to PA recommendations, the quality of the physician/patient relationship, illness-related beliefs, and demographic and personal information among adults who have LQTS? Specifically, the study explored and described the following: (a) frequency of participation in the PAs; (b) activities from the recommended guidelines that adults with LQTS believe to be permitted, not permitted, or about which clarity is lacking; (c) changes in the frequency of participation in the PAs since being diagnosed with LQTS; (d) adherence to PA recommendations, as indicated by participation in activities that are not permitted by their physicians; (e) and reported importance of participation in PAs. In addition, what PAs are adults with LQTS participating in that are not included on the recommended guidelines?

Furthermore, are factors associated with the quality of the physician/patient relationship, such as general communication, adherence dialogue, patient satisfaction, and working alliance related to illness-related beliefs, including treatment control, personal control, and illness coherence? Do ratings of adherence dialogue, patient satisfaction, communication, and working alliance predict levels of treatment control and personal control, respectively?

Primary Hypotheses

Hypothesis 1: Ratings of adherence dialogue (as measured by the Adherence Dialogue Scale), patient satisfaction (as measured by the Overall Satisfaction with Care Scale), communication (as measured by the Overall Communication Scale), and working

alliance (as measured by the Working Alliance Inventory Short-Client Form) would predict treatment control (as measured by the Illness Perception Questionnaire subscale).

Hypothesis 2: Ratings of adherence dialogue (as measured by the Adherence Dialogue Scale), patient satisfaction (as measured by the Overall Satisfaction with Care Scale), communication (as measured by the Overall Communication Scale), and working alliance (as measured by the Working Alliance Inventory Short-Client Form) would predict personal control (as measured by the Illness Perception Questionnaire subscale).

Post hoc Hypotheses

Post hoc hypothesis 1: Change in PA participation before and after LQTS diagnosis would be predicted by the quality of the physician/patient relationship, as measured by patient satisfaction, adherence dialogue, general communication, and working alliance, as well as by illness-related beliefs, including treatment control, personal control, and illness coherence. A multiple regression analysis was used to evaluate this hypothesis.

Post hoc hypothesis 2: Nonadherence, as indicated by participation in activities that are not permitted, would be predicted by patient satisfaction, adherence dialogue, general communication, and working alliance, as well as by treatment control, personal control, and illness coherence. This hypothesis was assessed using a logistic regression.

Chapter 3

Method

Research Design and Justification

Due to the risk of cardiac arrest associated with LQTS, along with the challenging psychosocial issues that arise, biopsychosocial research focusing on individuals with LQTS has increased. Although research with this population is advancing, many questions remain unanswered. LQTS is a complex and individualized condition, and information regarding risk stratification currently includes general population characteristics, such as gender, age, and type of LQTS (Priori et al., 2003). Consequently, the guidelines provided by the 36th Bethesda Conference included general recommendations for physicians to follow when treating patients with LQTS, but physicians must use specific information about the patient to tailor treatment recommendations regarding PA (Johnson & Ackerman, 2013; Roston et al., 2013). Therefore, recommendations regarding PA for the LQTS population are likely to be highly variable. Due to the high variability in the population and the need for further descriptive research concerning adherence to PA recommendations, a descriptive, self-report, survey design was used for the present study.

The purpose of the study was to describe and explain the physician/patient relationship, illness beliefs, and adherence in adults with LQTS by conducting a survey. The variables of interest were assessed by incorporating measures in an Internet survey format to enhance the validity and replicability of the study (Chris & Stewart, 2000). Web-page-based surveys offer the benefits of appearing identical to all participants, being able to reach many people, and obtaining data in a consistent fashion

(Chris & Stewart, 2000). In addition, an Internet survey offers participants anonymity, which is important when reporting sensitive information (Chris & Stewart, 2000), such as non-adherent behaviors. A potential disadvantage of using this research design is the perception as unwelcome or intruding on participants through Internet recruitment (Chris & Stewart, 2000). To address this possible issue, recruitment letters informed participants of the positive nature and objectives of the study (Appendix A). Online recruitment may also exclude individuals who avoid thinking about LQTS as a coping strategy and may not participate on such forums.

Participant Selection

All participants were 18 years or older, were formally diagnosed with LQTS (per participant report), and had Internet access. A total of 276 participants were recruited. Of these, 124 completed the personal questionnaire and PA survey, and 91 completed the entire survey. A minimum of 84 participants were needed for an alpha of.05 and medium effect size when measuring four independent variables (Cohen, 1992). Of the 91 participants, seven participated in other studies conducted by the Philadelphia College of Osteopathic Medicine.

Recruitment. Because the LQTS population is relatively small, and an appropriate sample size was unlikely to be acquired through traditional recruitment in medical facilities for the present study, participants were recruited via Internet resources. Two Internet resources that have high traffic volumes are Craigslist and Facebook. These sites, along with the Sudden Arrhythmia Death Syndromes website, Parent Heart Watch, Twitter, and other social media and LQTS/cardiac sites, were used. Additionally, the principal investigator is part of the LQTS research team at the

Philadelphia College of Osteopathic Medicine and has access to contact information for individuals who have participated in previous LQTS studies. These individuals were also contacted via e-mail with a recruitment letter.

Inclusion criteria. Participants met the criteria of being at least 18 years old, were residents of the United States, and were self-reportedly diagnosed with LQTS. In addition, participants were proficient in reading and understanding English and had access to the Internet. Individuals of all races and ethnicities who met the above criteria were able to participate.

Procedure

Data collection. Data was collected between January 2015 and June 2015. Adults with LQTS were recruited through Craigslist, Facebook, Twitter, and other social media and LQTS and cardiac websites. There were 27 LQTS support groups on Facebook with a range of five members to more than 900 members. All groups that had "LQTS" or "Long QT Syndrome" in the group title were contacted. Facebook group leaders were then contacted to obtain permission to post a recruitment letter on the group page (Appendix A). Once permissions were obtained, a recruitment letter that included an explanation of the study and contact information was submitted on the Facebook LQTS group pages (Appendix A). In addition, recruitment letters containing a link for the survey were posted to North American cities on Craigslist every 3 days. LQTS and cardiac websites and groups whose administrators gave the experimenter permission to recruit received a letter with information pertaining to the study and a link to the survey.

The survey used to obtain data was generated through Survey Monkey, an online resource used to create and administer surveys. After participants clicked on the link to

the survey, they were directed to the cover letter and were asked to click an area indicating that they reviewed and agreed with the information provided. Participants were prompted to complete and submit the online survey. Each survey included the adherence measure created for the study (Appendix B), the IPQ-R (Moss-Morris et al., 2002) the Satisfaction with Physician Scale (Davies & Ware, 2001), Adherence Dialogue Scale (Schneider, Kaplan, Greenfield, Li, & Wilson, 2004), General Communication Scale (Wilson & Kaplan, 2000), Working Alliance Inventory Short Client Form (Tracey & Kokotovic, 1989), and a personal information questionnaire. The survey required approximately 20 minutes to complete. Additionally, resources were included on the survey, including links to information pertaining to LQTS, depression, and anxiety.

After the survey was completed, participants were given the option to enter a lottery for a 1:10 chance of winning a $10 gift card from Walmart or Target. If participants chose to enter the lottery, they were asked to provide an e-mail address that would be used to contact the winner. Of 91 participants, 19 submitted an e-mail address to participate in the lottery. Two lottery winners were drawn.

Measures

Adherence. Adherence was measured by using the published recommended guidelines for athletic participation for individuals who have LQTS (Appendix B; Maron et al., 2004; Zipes et al., 2005). These were the most recent guidelines available during recruitment (January 2015 - June 2015). The guidelines include 28 specific activities of high intensity, moderate intensity, and low intensity activities, and participants were given the opportunity to list their top three PAs not included in the activities list. Participants were also asked to answer three questions regarding each activity to

determine adherence. Initially, they used a 7-point Likert scale consisting of *daily*, *weekly*, *monthly*, *bimonthly* (once every two months), *biannually* (once every 6 months), *annually*, and *never*, to report the frequency of participation in each activity since being formally diagnosed with LQTS by a physician. This method of scoring allowed adherence to be viewed on a continuum. Participants were then asked to report their cardiologists' recommendations for each activity by stating that the activity was recommended or not recommended or they were unsure if it was recommended. Additionally, participants were asked if they participated more, participated less, or there was no change in participation in the listed activities since being diagnosed with LQTS. Along with the questions specific to the activity list, participants were asked to report how important they believe PA is in general on a 7-point Likert scale indicating *not at all important*, *low importance*, *slightly important*, *neutral*, *moderately important*, *very important*, *extremely important* (Vagias, 2006). Additionally, information was obtained for post hoc analysis regarding participants' frequency of engaging in other behaviors that are typically recommended by cardiologists subsequent to a LQTS diagnosis. These activities include avoiding certain foods, medications, or substances that have been shown to result in an elongated QT interval, as well as drinking water, taking medications for LQTS, wearing a LQTS identification item, and taking a defibrillator on outings. Furthermore, participants who indicated that they had changed their participation in PAs before and after the LQTS diagnosis and were unsure if the activities are permitted by their physician were asked to explain why the change in participation occurred.

Quality of the physician/patient relationship. Three previously tested scales were used to evaluate the quality of specific factors that have been shown to influence the physician/patient relationship. Participants reported their overall satisfaction with care (Davies & Ware, 2001; Schneider et al., 2004), quality of adherence dialogue (Schneider, Kaplan, Greenfield, Li, & Wilson, 2004), and general communication with their cardiologists (Schneider et al., 2004; Wilson & Kaplan, 2000). All scales were completed by using a 5-point Likert scale: *excellent, very good, good, fair, or poor*.

To measure overall satisfaction with care, participants were asked to rate their cardiologist on personal manner, communication skills, technical skills, and overall care. The quality of adherence dialogue scale was developed for a previous study that examined the quality of the physician/patient relationship and adherence among patients diagnosed with HIV (Schneider et al., 2004). This scale was used in conjunction with the overall satisfaction with care scale and overall communication scale and for the Schneider et al. study. For the purposes of the present study, the word *HIV* was changed to *LQTS* and *antiretroviral medications* was changed to *PA recommendations* for the adherence dialogue scale. The authors of the scale recommended making these changes to reflect the medical population of interest.

Validity. Each scale has been demonstrated to be a valid tool for assessing important components of the physician/patient relationship. The authors who developed the general communication scale demonstrated that it has convergent and discriminant validity by evaluating the cross-sectional relationships of the scale with two measures of general satisfaction with care ($r = 0.85$, $p < .0001$; $r = 0.61$, $p < .0001$) and health-related quality of life ($r = 0.18$, $p < .005$; $r = 0.06$, $p < 33$; Wilson & Kaplan, 2000). The overall

satisfaction with care scale was published by the Group Health Association of America (Davies & Ware 2001). Additionally, the adherence dialogue scale was found to have convergent validity with satisfaction with care and overall communication (Schneider et a., 2004).

Furthermore, a study evaluated the quality of the physician/patient relationship using all three scales in 744 adults with HIV (Schneider et al., 2004). A principal components analysis demonstrated internal consistency, as Cronbach's alphas ranged from .92 to .93. Additionally, ordinal logistic regressions were conducted by using an odds ratio that represented the odds of changing by 1 level in a 20-level ordinal adherence variable for each unit of change in the independent variables. General communication was found to have an odds ratio of 1.15 (p = .0001), meaning that for each 10-point improvement in general communication, the odds of adherence improvement increased by 15%. Similarly, significant odds ratios were found between adherence to HIV medical recommendations and adherence dialogue (OR, 1.20, $p < .0001$) and overall satisfaction with care (OR, 1.14 , $p = < .0001$).

Working Alliance Inventory Short-Client Form (WAI-C). The WAI-C consists of 12 items that assess the strength of the working alliance from the client's or patient's perspective (Hatcher & Gillaspy, 2005; Tracey & Kokotovic, 1989). The inventory consists of three subscales that include agreement on treatment tasks, agreement on treatment goals, and the patient-provider emotional bond (Hatcher & Gillaspy, 2005; Tracey & Kokotovic, 1989). All 12 items were reworded to pertain to the medical relationship, and participants responded to the items using a 7-point Likert scale

(1, *strongly disagree*; 7, *strongly agree*). A sample item from the agreement on treatment goals subscale is "My doctor and I agree on my treatment plan." A sample item from the agreement on treatment tasks subscale is "My doctor and I agree about the things I need to do to help improve my health." A sample item from the emotional bond subscale is "My doctor understands all of what I am going through with my medical problem."

Validity and Reliability. The WAI-C has demonstrated high validity and reliability when used with medical populations (Fuertes et al., 2007). Specifically, the inventory has demonstrated excellent overall internal consistency, with a reliability alpha coefficient of .98 (Tracey & Kokotovic, 1989). Internal consistency has also been demonstrated among the tasks, goals, and emotional bond subscales independently, with alpha coefficients of .82, .72, and .98, respectively (Tracey & Kokotovic, 1989). Additionally, analysis of the correlations among the three subscales showed significant intercorrelations, ranging from .75 to .80 (Tracey & Kokotovic, 1989). A principal components analysis of all 12 items yielded in coefficient values ranging from .62 to .86 (Tracey & Kokotovic, 1989). Furthermore, the WAI is highly correlated with other measures evaluating working alliance, including the Penn Helping Alliance Scale and the California Psychotherapy Alliance Scale (Tichenor & Hill, 1989; Marmar et al., 1989).

Illness Perception Questionnaire-Revised (IPQR-R). The IPQ-R (Moss-Morris et al., 2002) was developed to evaluate the components that comprise the self-regulatory model (Leventhal et al.,1992) and has been shown to be appropriate for both male and female adult patients with cardiac diseases (Moss-Morris et al., 2002). The scale is comprised of seven factors: timeline of illness, illness changeability, illness

consequences, treatment control, personal control, emotional representation, illness coherence, and illness representation. These components provide a framework for patients to understand their illness and health risk that guides action, such as coping and adherence to treatment (Moss-Morris et al., 2002). Although all seven components comprise individuals' illness representation, three specific subscales have been shown to influence adherence levels to medical recommendations (Moss-Morris et al., 2002): treatment control, personal control, and coherence of one's illness.

Treatment control, personal control, and illness coherence subscales. The treatment control, personal control, and illness coherence subscales were used for the current study. These subscales contain a 5-point Likert scale consisting of *strongly disagree, disagree, neither agree nor disagree, agree,* and *strongly agree* that participants use to respond to each item.

Treatment control and personal control were divided into two separate subscales for the revised IPQ, after a factor analysis demonstrated that all items pertaining to control loaded on two separate dimensions (Moss-Morris et al., 2002). Specifically, the treatment control subscale includes four items that assess patients' beliefs regarding their treatment and recommended medical advice and has an alpha of .80 (Moss-Morris et al., 2002). The factor analysis also revealed that items loading on the personal control subscale reflect one's beliefs pertaining to their personal control over the illness and sense of self-efficacy (Moss-Morris et al., 2002). This subscale has six items and an alpha of .81 (Moss-Morris et al., 2002).

The illness coherence subscale was also added to the revised IPQ to evaluate the extent to which a medical patient's illness representation provides a coherent

understanding of the illness. Items in this subscale evaluate a type of metacognition that occurs when patients evaluate the coherence or usefulness of their illness representations (Moss-Morris et al., 2002). There are five items in this subscale, which has been demonstrated to have an alpha of .87 (Moss-Morris et al., 2002).

Validity and reliability. The validity and reliability of the IPQ-R have been demonstrated through extensive research in patients of various medical populations (Moss-Morris et al., 2002). High structural validity and internal reliability were demonstrated after two principal components analyses were conducted; alpha scores for the subscales ranged from .79 to .89. A 6-month evaluation of test-retest reliability conducted with two different groups of medical patients demonstrated stable correlations ranging from .46 to .88. Additionally, discriminant validity for the instrument was confirmed by using Pearson's correlations between subscales of the IPQ-R and The Positive and Negative Affect Schedule (Watson, Clark, & Tellegen, 1988). Researchers who revised the instrument also identified high predictive validity with medical patients by using data retrieved from a sample with multiple sclerosis (Moss-Morris et al., 2002). Furthermore, qualitative studies have provided evidence for the interrater reliability of each of the components of Leventhal's model (Weinman, Petrie, Sharpe, & Walker, 2000).

Demographic and personal information. In addition to questions pertaining to adherence, the physician/patient relationship, and illness beliefs, personal and demographic information was obtained (Appendix C). Studies have shown that factors including soceioeconomic status (Zolnierek, Haskard, & Dimatteo, 2009), race concordance of patient and physician (Cooper et al., 2003), gender (Turner et al., 2003),

and age (Schneider et al., 2004) may impact the physician/patient relationship and medical adherence. These characteristics, along with religion, were obtained from participants for the present study, along with LQTS-specific information.

Chapter 4

Results

Statistical Package for the Social Sciences software, Version 22 (IBM Corp, 2013), was used to conduct the analyses. Initial analysis began with examination of participants' characteristics assessed using the personal information questionnaire (PIQ; Appendix C), adherence characteristics (Appendix B), the physician/patient relationship, and LQTS-related beliefs. The descriptive review was followed by testing of hypotheses and post hoc analyses. The PIQ incorporated questions pertaining to participants' demographics, general medical information, family characteristics, LQTS symptom history, and LQTS treatment characteristics.

Initial Exploratory Findings

PIQ: demographic information. Demographic information was obtained using the PIQ pertaining to family history, medical history, LQTS symptoms, and LQTS treatment. Variability of demographic factors pertaining to the 91 participants who completed the entire survey was identified with regard to age, income, work status, education, and geographic location, while less variability was present with regard to gender, ethnicity, and religious beliefs (Table 1). These findings are similar to results obtained from the 124 participants who did not complete the survey beyond the PIQ and first PA list. Therefore, analysis was conducted using data collected from the 91 participants who completed the entire survey, as this group is likely to be representative.

Table 1

Demographic Variables

	Complete	**Partial**
Variable	***n* = 91**	***n* = 124**
Gender		
Male	3	5
Female	88 `	119
Age		
Range	49	53
Mean (SD)	39.55 (12.31)	40.24 (11.78)
Income		
$5,000-$20,000	5	6
$21,000-$40,000	17	20
$41,000-$60,000	10	16
$61,000-$80,000	13	13
$81,000-$100,000	7	14
Above $100,000	21	34
Rather Not Say	18	21
Work status		
Employed Full Time	35	49
Employed Part Time	16	21
Homemaker	16	22
Unemployed	9	9

(continued)

Variable	Complete *n* = 91	Partial *n* = 124
Work status		
Student	6	3
On Disability	5	9
Retired	4	5
School status		
Out of School	79	108
In School	12	16
Education		
Bachelor's Degree	28	36
Associate's Degree	23	32
High School	18	26
Master's Degree	10	15
Doctoral Degree	3	4
Other:	9	11
Did not graduate from high school, college, or completed trade school		
Ethnicity		
White	85	118
Hispanic	3	3

(continued)

Variable	Complete *n* = 91	Partial *n* = 124
Ethnicity		
Black	2	2
Asian	1	1
Residential area		
Suburban	44	63
City	28	37
Rural	13	17
Farm	3	4
Island	2	2
Region		
Northeast	32	40
South	23	33
Midwest	21	33
West	15	18
Religion		
Christian	73	94
Agnostic	6	11
Atheist	3	3
Hindu	1	1
Muslim	1	1

Note. Complete = participant who completed the entire survey; *n* = 124; Partial = participants who completed the PIQ and PA measures, but did not complete the physician/patient relationship question or the IPQ-R questions.

PIQ: LQTS and other general medical information. In addition to demographic information, participants were asked to report the age that they were first diagnosed with LQTS, if they had genetic testing, and LQTS type (Table 2). If participants endorsed medical conditions in addition to LQTS, they were prompted to list the conditions. Fifty participants reported that they had medical conditions in addition to LQTS. Conditions included cardiovascular, mental health, genetic, autoimmune, neurological, oncological, gynecological, and other chronic conditions. Of these, the most frequently reported conditions were cardiovascular, chronic health, and mental health conditions. Cardiovascular conditions included atrial fibrillation, leaky heart valve, "blood clotting disorder," myocardial infarction, hypertension, and cardiomyopathy. Chronic health conditions described by participants included diabetes, arthritis, sleep apnea, hyperlipidemia, anemia, asthma, thyroid disease, and gastroesophageal reflux disease. The most frequent mental health difficulties were anxiety (social and PTSD) and depression.

Table 2

LQTS Information

Variable	***n* = 91**
Age First Diagnosed	
Range	10-59
Mean	29.52
Modes	14,16
Genetic Testing	
Yes	77
No	14
Type of LQTS	
LQTS 1	30
LQTS 2	31
LQTS 3	6
LQTS 4	2
LQTS 6	2
LQTS 1 and 3	1
LQTS 1, 2, and 3	1
Waiting on results	2
Unknown	17

Along with diagnosed medical conditions, participants were asked if they suspected any undiagnosed medical conditions. Of the 91 participants, 79 indicated that

they do not suspect an undiagnosed condition. The remaining 12 participants reported various suspected illnesses, including mental health (anxiety, attention deficit disorder), immunological, oncological, and vascular conditions. Participants were also asked if diagnosed and/or suspected medical conditions impacted their participation in PAs. Most participants who endorsed an impact on PA (n = 25) stated that LQTS or LQTS-related factors (e.g. ICD and cardiac arrest) contributed to changes in participation. The second most frequently reported conditions for changing activity level involved pain (arthritis, spine injuries, and spine malformations).

PIQ: family information. Along with general medical information, participants also commented on pertinent family characteristics. Information regarding household members, number of children, and number of children with LQTS, whether children and parents see the same cardiologist, other members in the family have LQTS, and family members were symptomatic was obtained (Table 3). The majority of participants lived with a spouse/partner and had children. Of the 71 children reported, 51 had been diagnosed with LQTS, and most did not see the same cardiologist as their parent.

Table 3

Family Information (n = 91)

Variable	Frequency	% of Sample
Household		
Parents	4	
Siblings	1	
Roommate(s)	3	
Spouse/Partner	75	
Children	61	
Lives Alone	5	
Other	2	
Children		
0	14	15
1	16	18
2	24	26
3	20	22
4	10	11
5	1	1

(continued)

Variable	Frequency	% of Sample
Children with LQTS		
0	14	15
1	28	31
2	19	21
3	3	3
4	1	1
Unknown	7	8
Children see same cardiologist		
Yes	11	
No	50	
Other members in family have LQTS		
Yes	73	
No	18	
Asymptomatic family members		
Yes	56	
No	14	
Unsure	21	

PIQ: LQTS symptom history. In addition to general medical information, participants were asked to answer specific questions about their LQTS symptoms. Among the 91 participants who completed the entire survey, 70 reported that they are symptomatic which may include cardiac arrest, seizures, syncope, and palpitations. These participants were asked to indicate their age when the first LQTS symptom was experienced, the setting during the first symptom, if the first symptom required an emergency room visit, the month and year of the most recent symptom, frequency of symptoms, and the total number of symptomatic events. In addition, type of symptoms experienced were assessed as part of the IPQ-R and are listed below (Table 4).

Table 4

LQTS Symptom History (n = 70)

Variable	**Frequency**
Age during first symptom	
Range	57
Mean	18.41
Setting during first symptom	
Home	30
PA	21
School	10
Other	6
Work	4
Emergency room visit during first symptom	
Yes	39
No	28
Most recent symptom	
2015	15
2014	12

(continued)

Variable	Frequency
Most recent symptom	
2008-2013	6
2002-2007	7
1996-2001	3
1985-1995	1
1974-1984	2
Unknown	4
Symptom frequency	
Weekly	10
Monthly	10
Yearly	21
1-3 times	14
Never	15
Total number of symptoms	
0	2
1 or 2	17
3 or 4	14

(continued)

Variable	Frequency
Total number of symptoms	
5 or 6	11
7 or more	26
Percentage of symptom type *(n = 91)*	
Cardiac arrest	39
Syncope	67
Seizures	29
Heart palpitations	90
Dizziness	8

PIQ: LQTS treatment history. Information pertaining to LQTS treatment characteristics was also elicited from participants. These questions assessed presence of ICD/pacemaker, medications and side effects, and information pertaining to LQTS-related medical visits. Among the 91 participants, 36 did not have an ICD or pacemaker, 29 had both an ICD and pacemaker, 25 had an ICD alone, and one had a pacemaker alone. Eighteen participants reported that they own an AED and described when they carried the device with them. The vast majority reported that their decision to take an AED on outings was determined by the presence of their children who also have

LQTS. Other responses included that participants leave the AED in the car, take it when they expect that the destination does not have an AED, and take it to amusement parks.

Along with surgical treatment and AED use, information regarding pharmacological treatment was also elicited. Participants were first asked to list all medications they were currently taking. The most common medications reported were beta blockers. The remaining medications were variable and consistent with common chronic medical conditions in the United States (e.g. hypertension, hyperlipidemia, thyroid disease, diabetes, and pain). Side effects from LQTS medications were reported by 62 of the 91 participants. The most common side effects reported were fatigue, dizziness, hypoglycemia, low blood pressure, hair loss, and weight gain.

As part of the treatment history portion of the PIQ, information pertaining to LQTS-related medical visits was also obtained. The initial portion of these questions assessed if participants had changed cardiologists, frequency of visits, and date of most recent visit (Table 5). Information on other professionals that participants discussed LQTS with and frequency of discussing LQTS was also elicited. Most participants reported that they discuss LQTS with all doctors, general practitioners, obstetricians and gynecologists, and dentists. The presence of LQTS discussions with non-cardiology professionals varied from *never* to *during every visit* and *when needed*. The date of participants' last scheduled cardiology visit ranged from June 2012 to May 2015. Nearly half (45) of the reported visits were between October 2014 and March 2015.

Table 5

Treatment History

Variable	***n* = 91**
Changed cardiologists	
Yes	50
No	41
Total number of cardiologists visited	
1	7
2	27
3	12
4	4
5	1
6	2
7	1
NA	2
Frequency of cardiologist visits	
Every 3 months	11
Every 4 months	13

(continued)

Variable	***n* = 91**
Frequency of cardiologist visits	
Once/year	36
Twice/year	36
Other	25

The final portion of the treatment history section of the PIQ included questions pertaining to emotional difficulties. Among the 91 participants, 44 reported that they had sought help from a healthcare professional for emotional issues. The top conditions specified by participants that had resulted in seeking help were anxiety and depression. Along with treatment history, participants were asked to report what they thought was the most important factor to keep them safe and healthy. Many stated that having personal awareness of cardiac event triggers, awareness of the LQTS diagnosis, avoiding all cardiac event triggers (e.g., "physical activity, certain medications, stress, and poor overall health") and regular communication with physicians are the most important factors in regard to LQTS treatment. Others reported medications and their ICD to be the most important.

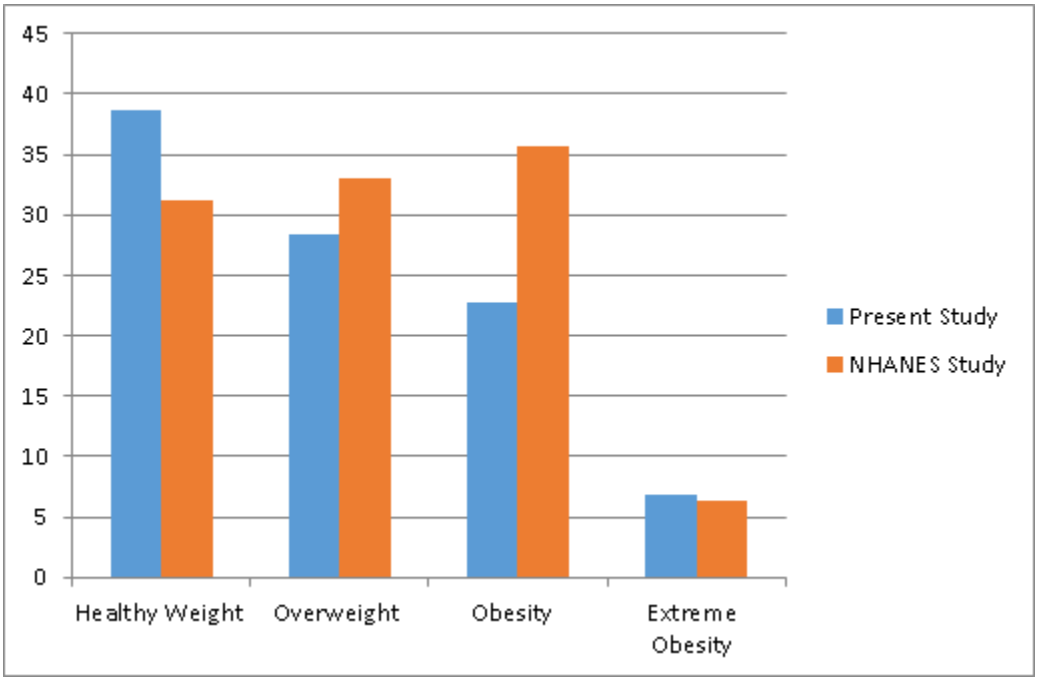
PA Participation: Descriptive Statistics

Descriptive statistics were used to analyze adherence characteristics, physician/patient relationship characteristics, and LQTS-related beliefs. The following

information pertinent to adherence to PA recommendations was obtained: (a) body mass index (BMI); (b) ratings of importance of participation in PAs; (c) total frequency of participation in PAs; (d) frequency of participation in the top three PAs listed by the participant; (e) change in frequency of participation in activities since being diagnosed with LQTS; (f) frequency of participation in the Bethesda Guidelines PAs list; (g) activities reported to be permitted, not permitted, or unclear whether the activities are permitted by their cardiologists, and (h) adherence to PAs that at least 10% of participants engaged in.

BMI is a measure of body fat based on height and weight and is used in the United States to determine if an individual is of low weight (18.5), normal weight (18.5 to 24.9), overweight (25.0 to 29.9), obese (30 or more), or extremely obese (40 or more). The three men who participated in the study reported BMIs of 81.8, 41.84, and 22.43. The men were not included in the BMI analysis in order to compare the majority of the participants, who were women, to the most accurate normative data. Figure 1 compares BMI percentages in each category among participants in the present study to data for adult women in the United States in 2009 collected by the National Health and Nutrition Survey (NHANES, 2009).

Figure 1. Comparison of BMI Between Present Study and NHANES Survey (%)



Importance of PA was measured by a 6-point Likert scale. The number of participants who chose each rating varied from low importance (7), slightly important (10), neutral (13), moderately important (18), very important (25), to extremely important (18). Frequency of participation in PAs was also recorded by calculating the total number of participants who engaged in PA at various levels. Most participants engaged in exercise on a weekly (38) or daily (37) basis. The remaining participants reporting that they never participated in PAs (12) or they participated on a monthly (2) or biannual (2) basis.

To account for activities that are not listed in the Bethesda Guidelines (Appendix B) that individuals may engage in regularly, participants were prompted to list the top three PAs that they participated in the most before being shown the guidelines. Many participants reported that their top PA was walking (42), while other cardiovascular

exercises (running, jogging, and swimming) and weight training were also reported frequently. There was high variability among the second and third top PAs, with many reports of walking, weight training, household chores, and caring for children.

Due to PA restrictions recommended for individuals with LQTS (Zipes et al., 2005), change in PA participation after receiving a LQTS diagnosis was assessed with a 5-point Likert scale that ranged from 1 equals not at all to 5 equals very much. Twenty-nine participants reported that PA changed very much, 21 reported no change in PA, and 21 endorsed moderate change.

The Bethesda Guidelines are a list of PAs that should be permitted, permitted on an individual basis, or not permitted by physicians who treat LQTS (Zipes et al., 2005). Cumulative percentages of participation in each activity along with number of participants with an implantable device, were recorded (Table 6).

Table 6

Participation (%) in Physical Activities Published by the Bethesda Guidelines

High Intensity Activities					
Not Permitted		**Permitted on Individual Basis**		**Permitted**	
% Participation (% with an implantable device)					
Running	33.3 (80)	Soccer	7.8 (86)		
Basketball	12.2 (55)	Skiing	6.7 (83)		
Tennis	7.9 (57)	Racquetball/ Squash	0 (0)		
Rock climbing	10 (40)				
Body building	7.8 (42)				
Touch football	4.4 (75)				
Windsurfing	0 (0)				
Ice hockey	0 (0)				
Moderate Intensity Activities					
% Participation (% with an implantable device)					
Swimming	66.3 (61)	Hiking	56.2 (54)	Treadmill	68.9 (56)
Weightlifting	40.4 (50)	Jogging	41.6 (59)	Biking	48.9 (61)
Motorcycling	7.9 (29)	Baseball/Softball	12.2 (90)		
Surfing	3.4 (33)	Sailing	3.4 (100)		
Low Intensity Activities					
% Participation (% with an implantable device)					
Snorkeling	13.6 (67)	Horseback riding	18.9(71)	Brisk walking	88.9 (55)
Scuba diving	4.4 (50)			Bowling	46.7(52)
				Skating	18.2 (44)
				Golf	14.4 (31)

Participants were given the Bethesda Guidelines PA list (Appendix B) a second time to indicate which activities were permitted, not permitted, or possibly permitted by their cardiologists. Table seven presents recommendations by participants' cardiologists in comparison to the Bethesda Guidelines. The percentage of participants who have engaged in each activity within the last year is also presented.

Table 7

Bethesda Guidelines, Physician Recommendations, and Participation in Physical Activities

Physician Recommendation (%)	***Permitted***	***Not Permitted***	***Unsure***	***Not Discussed***
	% Physicians' recommendation (% of participation)			
Bethesda Guidelines				
Permitted				
Biking	45.6 (36)	7.8 (2)	7.8 (3)	38.9 (55)
Treadmill	67.0 (48)	4.4 (1)	4.4 (2)	24.2 (13)
Bowling	40.9 (27)	4.5 (1)	10.2 (1)	44.3 (15)
Golf	32.6 (9)	5.6 (1)	9.0 (0)	52.8 (2)
Skating	21.3 (3)	9.0 (0)	10.1 (1)	59.6 (13)
Brisk walking	78.0 (73)	1.1 (1)	3.3 (2)	17.6 (14)
Permitted on Individual Basis				
Racquetball/	10.1 (1)	19.1 (1)	7.9 (0)	62.9 (0)
Squash				
Skiing	18.0 (3)	14.6 (0)	11.2 (2)	56.2 (2)
Soccer	20.2 (9)	19.1 (2)	5.6 (0)	55.1 (1)
Baseball/	24.7 (9)	18.0 (2)	6.7 (1)	50.6 (2)
Softball				
Jogging	43.8 (32)	13.5 (0)	9.0 (2)	33.7 (9)
Sailing	14.6 (1)	6.7 (0)	13.5 (1)	65.2 (2)
Hiking	52.2 (41)	8.9 (1)	8.9 (2)	30.0 (13)
Horseback riding	24.4 (8)	3.3 (0)	15.6 (1)	56.7 (10)
Not Permitted				
Basketball	18 (8)	25.8 (3)	3.4 (0)	52.8 (2)
Body building	14.4 (7)	20.0 (0)	7.8 (1)	57.8 (3)

Ice hockey	4.5 (0)	18.2 (1)	9.1 (0)	68.2 (0)
Rock climbing	14.6 (7)	15.7 (0)	9.0 (1)	60.7 (3)
Running	35.6 (27)	18.9 (0)	11.1 (2)	34.4 (5)
Tennis	16.9 (7)	15.7 (1)	10.1 (0)	57.3 (3)
Touch football	12.4 (3)	15.7 (0)	6.7 (0)	65.2 (2)
Windsurfing	6.7 (1)	14.6 (0)	7.9 (0)	70.8 (0)
Motorcycling	15.6 (8)	7.8 (0)	16.7 (3)	60.0 (1)
Surfing	8.0 (2)	15.9 (0)	11.4 (1)	62.8 (1)
Swimming	49.4 (36)	23.6 (13)	2.2 (2)	24.7 (12)
Weightlifting	37.8 (25)	13.3 (0)	11.1 (3)	37.8 (12)
Scuba diving	2.2 (0)	19.1 (0)	12.4 (1)	66.3 (2)
Snorkeling	13.8 (5)	10.3 (1)	13.8 (1)	62.1 (5)

Information on adherence to cardiologists' recommendations was obtained by presenting the Bethesda Guidelines list to participants twice. When the list was first presented, participants were asked to report frequency of participation in each activity during the last 12 months by choosing daily, weekly, monthly, bimonthly, biannually, annually, or never. When the list was presented the second time, participants were asked to report if each activity was permitted, not permitted, possibly permitted, or has not discussed with their cardiologist. For the purpose of this study, adherence was defined as any level of participation in an activity that at least 10% of individuals engaged in and was not permitted by their cardiologist. PAs that at least 10% of participants endorsed were basketball, body building, rock climbing, running, soccer, tennis, baseball/softball, biking, motorcycling, jogging, swimming, treadmill, weightlifting, hiking, bowling, golf, horseback riding, skating, snorkeling, and brisk walking. Table 8 shows the number of

participants who were adherent or non-adherent to recommendations for each of these activities.

Table 8

Number of Participants Adherent to Physical Activities

Activity	Adherent	Non-Adherent	Missing
Basketball	85	3	0
Body building	89	0	2
Rock climbing	87	0	4
Running	87	0	4
Soccer	86	2	3
Tennis	86	1	4
Baseball	86	2	3
Biking	88	1	2
Motorcycling	88	0	3
Swimming	75	12	4
Treadmill	89	1	1
Weightlifting	88	0	3
Hiking	87	1	3
Bowling	86	1	4
Golf	87	1	3
Horseback riding	88	0	3
Skating	87	0	4
Snorkeling	82	1	8
Brisk walking	90	0	1
Jogging	87	0	4

Physician/patient relationship. The physician/patient relationship was measured by adherence dialogue, overall satisfaction with physician, general communication, and working alliance. Working alliance was divided into three components: agreement on treatment task, agreement on treatment goal, and emotional bond. Table 9 shows how the participants rated these variables.

Table 9

Quality of the Physician/Patient Relationship Descriptive Statistics

Variable	Mean	Median	Standard Deviation
1 = excellent; 5 = poor			
Adherence dialogue	2.27	2.0	1.16
Overall satisfaction	1.83	1.0	1.15
General communication	2.08	1.6	1.2
1 = never; 7 = always			
Goal	3.9	4.0	.77
Task	5.14	5.5	1.5
Bond	5.58	6.0	1.46

LQTS beliefs. The IPQ-R (Moss-Morris et al., 2002) is comprised of seven factors that provide a framework for patients to understand their illness and health risk that guides action, such as adherence to treatment (Moss-Morris et al., 2002). Participants rated each subscale using a 5-point Likert scale (1 = strongly

disagree; 5 = strongly agree). Table 10 describes how the present sample rated all seven subscales.

Table 10

IPQ-R Subscale Descriptive Statistics

Variable	Mean	Median	Standard Deviation
Timeline Acute/Chronic*	23.78	25.00	3.26
Timeline cyclical*	10.34	11.00	4.1
Consequences*	21.76	22.00	4.82
Emotional Representation*	16.87	16.0	5.6
Personal Control**	21.16	21.00	5.1
Treatment Control**	15.77	16.00	3.70
Illness Coherence**	20.36	21.00	4.77

Note. *High ratings (Mean = 30) represent strongly held beliefs about the number of symptoms attributed to LQTS, the chronicity of the condition, the negative consequences of LQTS, the cyclical nature of the condition, and the emotional impact of LQTS.

**High ratings (Mean = 25) represent positive beliefs about the controllability of the illness and a personal understanding of the condition.

Physician/patient relationship and LQTS beliefs. Product moment Correlations were calculated between the physician/patient relationship and LQTS-related beliefs. To assess the physician/patient relationship, adherence dialogue, overall satisfaction with physician, general communication, and working alliance were evaluated. Illness perceptions were assessed with the IPQ-R (Moss-Morris et al., 2002) and included beliefs about timeline, consequences, personal control, treatment control, illness coherence, emotional representation, and identity. Perceptions about the

chronicity of LQTS were not correlated with other variables. Ratings of overall satisfaction with the physician was highly associated with all other physician/patient relationship variables ($p < .01$) and not significantly correlated with any perception variables. All additional findings are presented in Table 11.

Table 11

Product Moment Correlations Between Physician/Patient Relationship Variables and Illness Perceptions

	1	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**
1	1	.816**	.750**	.744**	-.347**	-.055	-.077	-.060	-.328**	.005	.000
2	.816**	1	.805**	-.770**	-.527**	-.062	-.213*	-.120	-.357**	-.074	.025
3	-.750**	.805**	1	.842**	.541**	.089	.187	.147	.289**	.099	.034
4	-.744**	.770**	.842**	1	.554**	-.065	.204	.199	.279**	.118	-.043
5	-.347**	.527**	.541**	.554**	1	-.058	.218*	.211*	.092	.203	-.096
6	-.055	-.062	.089	-.065	-.058	1	.022	-.322**	.142	.308**	.529**
7	-.077	-.213*	.187	.204	.218*	.022	1	.433**	.245*	-.133	-.179
8	-.060	-.120	.147	.199	.211*	-.322**	.433**	1	.039	-.026	-.392**
9	-.328**	.357**	.289**	.279**	.092	.142	.245*	.039	1	-.205	.014
10	.005	-.074	.099	.118	.203	.308**	-.133	-.026	-.205	1	.273**
11	.000	.025	.034	-.043	-.096	.529**	-.179	-.392**	.014	.273**	1

Note. High scores on treatment tasks and goals indicate negative perceptions.

1. Overall Communication
2. Adherence Dialogue
3. Task
4. Bond
5. Goal
6. Consequences
7. Personal Control
8. Treatment Control
9. Illness Coherence
10. Timeline Cyclical
11. Emotional Representation

*$p < .05$ (2-tailed)
**$p < .01$ (2-tailed)

Two primary hypotheses were evaluated using multiple regressions. Multiple regressions are used to predict one variable from multiple predictor variables (Howell, 2011). Because previous research has not been conducted to evaluate the relationships between the independent and dependent variables, stepwise or setwise regressions were not used. Assumptions of homoscedasticity, multicollinearity, and non-zero variance were tested prior to conducting the analyses. Both dependent variables, personal control and treatment control subscales from the IPQ-R (Moss-Morris et al., 2001), met all assumptions. The WAI-SC (Hatcher & Gillaspy, 2005; Tracey & Kokotovic, 1989) goal subscale is the only predictor variable that demonstrated a homogenous sample. The remaining predictor variables did not meet assumptions of homoscedasticity and normal distribution (Table12; Table 13).

Table 12

Measures of Central Tendency of Predictor Variables

	Overall Communication Scale	**Adherence Dialogue Scale**	**Overall Satisfaction Scale**	**Task**	**Bond**	**Goal**
Mean	50.0	50.0	50.0	5.2005	5.5549	3.9533
Median	45.1076	48.1953	43.0224	5.7500	6.0000	4.0000
Skewness	.921	.674	1.205	-.917	-1.130	.346
Standard Error of Skewness	.253	.254	.254	.253	.253	.253
Kurtosis	-.425	-.697	.191	.105	.552	2.187
Standard Error of Kurtosis	.500	.503	.503	.500	.500	.500

Table 13

Z-scores for Skewness and Kurtosis of Predictor Variables

	Overall Communication	**Adherence Dialogue**	**Overall Satisfaction**	**Task**	**Bond**	**Goal**
Z skewness	3.64***	2.65**	4.74***	3.62***	4.46***	1.37
Z kurtosis	-0.85	-1.38	0.38	0.21	1.10	4.37***

** p < .05*
***p < .01*
****p < .001*

A forward multiple regression was calculated to predict treatment control (as defined by the IPQ-R subscale) based on participants' ratings of adherence dialogue (as measured by the Adherence Dialogue Scale), patient satisfaction (as measured by the Overall Satisfaction with Care Scale), communication (as measured by the Overall Communication Scale), and working alliance (as measured by the WAI- SC). All predictors except for agreement on treatment goal did not enter into the equation. Agreement on treatment goal significantly predicted perception that medical treatment controls LQTS ($F(1, 89) = 4.121$, $p = .045$). The adjusted R^2 value was .035, indicating that about 4% of the variance in perception that medical treatment controls their LQTS was explained by the model.

Table 14

Analysis of Variance

Model	***SS***	***df***	***MS***	***F***	***P***
Regression	59.479	1	59.479	4.121	.045
Residual	1284.6000	89	14.434		
Total	1344.080	80			

A forward multiple regression analysis was also used to test personal control (as measured by the IPQ-R subscale) based on participants' ratings of adherence dialogue (as measured by the Adherence Dialogue Scale), patient satisfaction (as measured by the Overall Satisfaction with Care Scale), communication (as measured by the Overall Communication Scale), and working alliance (as measured by the WAI-SC). Agreement on treatment goal was kept in the regression and was found to significantly predict perception of personal control over LQTS ($F(1,89) = 4.229$, $p = .043$). The adjusted R^2 value was .034, indicating that about 3% of the variance of perception of personal control over their LQTS is accounted for by the model.

Table 15

Analysis of Variance

Model	***SS***	***df***	***MS***	***F***	***P***
Regression	108.731	1	108.731	4.229	.043
Residual	2288.257	89	25.711		
Total	2396.989	90			

Post Hoc Analyses

Post hoc analyses were conducted to further assess adherence, quality of the physician/patient relationship, and illness-related beliefs. To continue to describe the exploratory data, chi-square and crosstabs were conducted. Medical recommendations, in addition to physical activity recommendations, were also described. Furthermore, two post hoc analyses were implemented.

Chi-square and cross-tabulation tests. Cross-tabulation and chi-square analysis are often used for descriptive survey studies (Aiken, 1997). Results demonstrated

sufficient variability in the data, suggesting that at least 10 people were non-adherent to recommendations on PA. Specifically, 12 participants reported that they were nonadherent to recommendations regarding swimming. The expected frequencies in each cell must be greater than 5 in order to assume the sampling distribution is close enough to a perfect chi square distribution. This assumption was not met due to the small sample size, and cross-tabulations were not conducted.

Cross-tabulation and chi-square analyses were used to determine if demographic variables and LQTS-related variables were statistically independent or if they were associated. Prior to conducting of a chi-square analyses, specific assumptions were tested. Data incorporated in the chi-square tests were categorical, the variables consisted of independent groups, and each cell in the cross-tabulation table had a value of at least 5. These tests were used to describe characteristics pertaining to adherence, participants who had changed cardiologists, those who had experienced various LQTS-related symptoms, and those who had sought help for emotional problems.

A total of 54.9% of participants reported that they had changed their cardiologist since being diagnosed with LQTS. Chi-square analyses were conducted to determine if a relationship exists between those who had changed their cardiologist and those who had symptoms of LQTS (76.9%) and those who had sought help for emotional problems (48.4%). A significant association was not found between change in cardiologist and experience of symptoms, x^2 (1) = .592. Similarly, a relationship was not identified between participants who changed their cardiologist and those who had emotional problems, x^2 (1) = 1.41.

A total of 48.4% of participants reported that they had sought help from a medical or mental health professional for emotional problems, such as anxiety and depression; 68.1% of participants reported experiencing side effects from medications. A chi-square analysis was conducted to evaluate if seeking help for emotional problems was associated with experiencing side effects from LQTS medications. A relationship was not found between these variables, x^2 (1) = .212. Chi-square analyses were also conducted to evaluate relationships between seeking emotional problems and specific symptom experience. There was no association between seeking help for emotional problems and cardiac arrest, x^2 (1) = .518, seizures, x^2 (1) = 2.78, and fainting, x^2 (1) = 1.25. Conversely, a significant relationship was present between seeking help for emotional issues and heart palpitations x^2 (1) = 5.55, $p < .05$. Based on the phi statistic, this association has medium strength (-.247) and is significant at $p < .019$.

Additional medical recommendations. Along with recording characteristics pertaining to common PA treatment recommendations, data related to other empirically supported medical recommendations for people with LQTS (Fitzgerald & Ackerman, 2005; Vyas, Johnson, Houlihan, Bauer, & Ackerman, 2006; Rottlaender, Motloch, Reda, Larbig, & Hoppe, 2012) was also obtained (Table 16). Over 50% of participants reported at least weekly participation in three of the six activities that are often recommended for LQTS (consuming potassium, sodium, and water). Activities that are recommended for individuals with LQTS that fewer than 50% of participants reported engaging in at least weekly were wearing a LQTS identification item, taking an AED on all outings, and drinking an electrolyte replacement drink. Of the activities that are not recommended for LQTS, 73% of participants reported daily to weekly caffeine consumption.

Table 16

Frequency of Participation Since LQTS Diagnosis

	Recommend	Daily	Weekly	Monthly	Bi Monthly	Bi Annually	Annually	Never
High Potassium Foods	Yes	52	25	5	1	5	0	5
High Sodium Foods	Yes	19	30	13	7	2	2	13
Grapefruit Juice	No	0	4	7	4	2	7	64
LQTS ID	Yes	25	2	2	2	1	1	53
AED	Yes	9	4	3	3	2	0	65
Caffeine	No	44	18	3	3	2	1	14
Substance Use	No	4	3	3	0	1	5	71
Alcohol	No	5	24	14	7	3	2	32
Cigarettes	No	8	1	0	0	2	3	72
Water	Yes	81	3	0	0	0	0	4
Electrolyte Drink	Yes	16	17	15	6	6	2	25

In addition to collecting frequencies of participation in the above activities, participants were asked to indicate which activities have been permitted, not permitted, or they are unsure if the activities are permitted by their cardiologists (Table 17).

Table 17

LQTS Recommendations, Physician Recommendations, and Frequency of Participation

		Physician Recommendation				
		Permitted	Not Permitted	Unsure	Not Discussed	% Participating
Recommended Activities						
Daily potassium		64	1	3	23	94.3
Daily sodium		23	16	9	41	84.9
LQTS ID		49	1	1	38	38.4
AED		24	7	4	55	24.4
Daily water		24	0	11	56	95.5
Electrolyte replacement drink		7	35	5	44	71.3
Discouraged Activities						
Grapefruit juice		11	27	6	46	27.3
Caffeine		31	22	10	27	83.5
Recreational drugs		8	33	5	44	18.4
Alcohol		31	16	8	33	63.2
Cigarettes		8	33	5	44	16.3

% equals cumulative participation in the activity

Along with common lifestyle modifications, adherence to PAs and other recommendations were identified and are presented in Table 18. Activities with the highest rates of nonadherence were swimming, consumption of sodium, and consumption of

caffeine. Overall, 59 participants reported adherence to all medical recommendations and 30 reported nonadherence.

Table 18

Adherence to All Recommendations

Activity	Adherent	Non Adherent	Missing
Basketball	85	3	0
Body building	89	0	2
Rock climbing	87	0	4
Running	87	0	4
Soccer	86	2	3
Tennis	86	1	4
Baseball	86	2	3
Biking	88	1	2
Motorcycling	88	0	3
Swimming	75	12	4
Treadmill	89	1	1
Weightlifting	88	0	3
Hiking	87	1	3
Bowling	86	1	4
Golf	87	1	3
Horseback riding	88	0	3
Skating	87	0	4
Snorkeling	82	1	8
Brisk walking	90	0	1
Jogging	87	0	4
Consuming potassium	89	1	1
Consuming sodium	76	12	3
Consuming grapefruit juice	86	1	4
Wearing LQTS ID	85	1	5

Consuming caffeine	79	11	1
Activity	**Adherent**	**Non Adherent**	**Missing**
Using AED	83	2	6
Using Substances	88	1	2
Consuming Alcohol	80	6	5
Smoking Cigarettes	86	3	2

Cross-tabulations and chi-square analyses were conducted to evaluate the relationship between nonadherence and symptom experience, given prior research indicating that symptom experience impacts adherence (Carney et al., 2010; Haynes & Sackett, 1979). A total of 77% of participants reported experiencing LQTS-related symptoms. A chi-square analysis was conducted to determine if a relationship existed between those with symptoms and adherence/nonadherence. Significant associations were not found between symptom experience and adherence x^2 (1) = .149. In addition to addressing symptom experience, illness perceptions about LQTS were also identified.

Illness identity comparisons. Beliefs developed about illness have been studied with the IPQ-R in various medical populations (Moss-Morris et al., 2002), including chronic/acute pain (Nicklas et al., 2010; Moss-Morris et al., 2002), hypertension (Ross, Walker & MacLeod 2004), rheumatoid arthritis (Moss-Morris et al., 2003), and multiple sclerosis (Bassi et al., 2015). Personality differences and illness perceptions among individuals with a history of myocardial infarction have also been assessed (Williams et al., 2010). Type D personality refers to the combined effects of negative affectivity and social inhibition as personality traits and has been shown to predict mortality in cardiac patients (Pederson & Denollet, 2006; Williams et al., 2010). Although the primary aim

of the study involved specific beliefs about personal control, treatment control, and illness coherence, four additional types of perceptions were also assessed. Illness perception findings using the IPQ-R and the Brief IPQ-R (Moss-Morris et al., 2002) among different medical populations, including the present study, are presented in Table 19.

Table 19

Sub Scale Comparisons among Different Populations (Mean (SD))

IPQ-R					**Brief IPQ**			
	Present Study	**Chronic Pain**	**RA***	**Acute Pain**	**Hypertension**	**MS****	**Myocardial Infarction**	
High scores (mean = 25) reflect positive beliefs					**High scores (mean = 10) reflect positive beliefs**			
							Type D	Non Type D
Personal control	21.16 (5.1)	18.42 (4.01)	19.97 (0.51)	22.94 (3.52)	3.49 (0.66)	3.03 (0.65)	5.67 (0.92)	6.39 (1.97)
Treatment control	15.77 (3.70)	19.43 (3.28)	16.78 (0.54)	19.43 (3.28)	3.53 (0.60)	3.7 (0.72)	6.43 (1.49)	7.79 (1.58)
Illness coherence	20.36 (4.77)	13.37 (4.78)	16.82 (0.61)	9.31 (3.00)	Not Reported	3.32 (0.77)	6.72 (0.65)	7.35 (1.21)
High scores (mean = 30) reflect a threatening view of illness					**High scores (mean = 10) reflect a threatening view of illness**			
Timeline - cyclical	10.34 (11.00)	9.37 (2.58)	13.82 (0.39)	9.37 (2.58)	3.21 (0.79)	3.46 (0.85)	7.92 (1.17)	6.43 (1.45)
Timeline – acute	23.78 (3.26)	13.40 (5.38)	23.38 (0.48)	13.40 (5.38)	3.55 (0.44)	4.30 (0.69)	7.92 (1.17)	6.43 (1.45)
Consequence	21.76 (4.82)	23.45 (3.89)	21.38 (0.56)	14.23 (4.44)	2.63 (0.63)	3.61 (0.76)	8.3 (0.9)	6.07 (1.11)
Emotion	16.87 (5.6)	19.75 (4.15)	15.92 (0.64)	16.12 (4.03)	3.01 (0.77)	3.19 (1.04)	7.58 (1.22)	5.64 (1.88)

Chronic Pain (Moss-Morris et al., 2002)
*Rheumatoid Arthritis (Moss-Morris et al., 2003)
Acute Pain (Moss-Morris et al., 2002)
Hypertension (Ross et al., 2004)
**Multiple Sclerosis (Bassi et al., 2015)
Myocardial Infarction (Williams et al., 2011)

The first post hoc hypothesis was that change in PA participation before and after LQTS diagnosis would be predicted by the quality of the physician/patient relationship, as measured by patient satisfaction, adherence dialogue, general communication, and working alliance, as well as illness-related beliefs, including treatment control, personal control, and illness coherence. Of all predictor variables, goal (WAI Subscale), personal control, and treatment control (IPQ-R subscales) met assumptions of heterogeneity and normal distribution. A multiple regression using the enter method was conducted with these three predictor variables, and a significant regression equation was not found (p = .5). This outcome should be interpreted with caution due to low variability displayed across dependent variable levels (1, *not at all*; 5, *very much*). A logistic regression was not implemented for comparison due to the inherent interpretation difficulties of creating artificial levels.

The second post hoc hypothesis was that nonadherence, as indicated by participation in activities that are not permitted, would be predicted by treatment control, personal control, and illness coherence. This hypothesis was tested using a binary logistic regression. Prior to conducting the analysis, assumptions of linearity, multicollinearity, and independence of errors were tested. Linearity was tested by evaluating the significance of the interaction between the predictor variables and their respective log transformations. Illness coherence failed to meet this requirement (p = .05). Interactions among the remaining independent variables were not significant and were kept in the regression. The multicollinearity assumption was also met with this data. The tolerance values were greater than .1 and VIF values were less than 10. Assumption of independence of errors was also met.

A forward stepwise logistic regression analysis was conducted on adherence to PA and lifestyle recommendations, using beliefs about personal control and treatment control as predictors. The Roa's score statistics for treatment control (p = .041) and the interaction of personal control and treatment control (p = .029) were significant. The personal control score alone was not significant (p = .136). The variable with the highest score statistic value, the interaction of treatment and personal control (x^2 (1) = 4.743, p = .029), was selected for inclusion in the model. Results are displayed in Table 20. High ratings of personal control indicated more negatively held beliefs and adherence was computed as 1 and nonadherence was computed as 0. As ratings of personal control and treatment control increased, the likelihood of adherence increased (Exp B = 1).

Table 20

Logistic Regression of Adherence, Treatment Control, and Personal Control

		95% CI for Odds Ratio		
	B(SE)	**Lower**	**Odds Ratio**	**Upper**
Included				
Constant	2.001 (.667)			
Treatment Control * Personal Control	-.004 (.002)	.993	.996	1.00

Chapter 5

Discussion

LQTS is a cardiac arrhythmia disorder that affects 1 in 2,000 individuals (Schwartz et al., 2016) and is a precursor to various cardiac events, including cardiac arrest, syncope, seizures, and heart palpitations (Vincent et al.,1999). Medical recommendations for LQTS often include pharmacological treatment (Yan & Kowey, 2011), surgical procedures (Dorostkar et al.,1999), and lifestyle modifications (Fitzgareld & Ackerman, 2005). PAs have been found to trigger cardiac events in individuals with LQTS (Aziz et al., 2011). Due to the seriousness of symptoms individuals with LQTS may experience when engaging in PA, adherence to the medical recommendations is important for survival (Yan & Kowey, 2011).

The quality of the physician/patient relationship and specific illness beliefs have been shown to predict adherence in various medical patient populations (Chen et al., 2011; Moss-Morris et al., 2001) but have not been researched with the LQTS population. The purpose of the present study was to explore the characteristics pertaining to adherence to PA recommendations, the quality of the physician/patient relationship, and illness beliefs in adults with LQTS. The quality of the physician/patient relationship was defined as overall communication, adherence dialogue, patient satisfaction, and working alliance. Specific illness-related beliefs that were assessed included patients' coherent understanding of the illness, as well as illness beliefs regarding personal control of the illness and treatment control.

A total of 276 participants were recruited. Of these, 91 completed the entire survey, and their data were used in the analyses. Demographic and LQTS-related

characteristics and adherence characteristics were identified and described as they related to the PA guidelines for LQTS from 2005 (Zipes et al., 2005), beliefs about personal and treatment control, and the new PA guidelines (Ackerman, Zipes, Kovacs, & Maron, 2015).

The PIQ collected participants' demographic information, general medical information, family characteristics, LQTS symptom history, and LQTS treatment characteristics. Variability of demographic factors was identified in regard to age, income, work status, education, and geographic location, while less variability was present in regard to gender and ethnicity. Of the 91 participants, 3 were men. Recruitment method and LQTS prevalence differences between genders may have contributed to this outcome. Internet social medial and support groups were used for recruitment. The most likely groups to utilize online health groups are White women between the ages of 18 and 49 with at least some college education who live in higher-income households (Fox, 2011). In comparison, the least likely populations to use online health groups are African Americans, Latinos, adults age 65 and older, adults with a high school education or less, and adults living in low-income households ($30,000 or less annual income; Fox, 2011). In addition to the inherent gender bias among people who use online health support groups (Fox, 2011), LQTS-specific factors may also contribute to the gender bias found in the present study. Women are diagnosed with inherited LQTS more often than men (Brenyo, Huang, & Aktas, 2012). This may be due to differences in the upper limit of the QT interval between post pubertal females and males (460 and 440 ms, respectively; Imboden et al., 2006). It is important to note that evidence obtained from other medical populations that suggests women are more likely to be medically non-

adherent than men (Lee & Khan, 2015; Lewey et al., 2013; Turner et al., 2003). In addition, a racial bias was also present. The majority of participants in the study were Caucasian. A recent study found that the prevalence among Caucasian individuals with LQTS is a 1:2534 ratio (Schwartz et al., 2016). Therefore, the racial composition in this study is likely to be representative of the LQTS population.

Participants also reported the age they were first diagnosed with LQTS, if they had genetic testing, and LQTS type. The youngest age of diagnosis was 10, indicating that no participants were diagnosed at birth, and families of participants may not have been aware that they were genetically predisposed to LQTS prior to diagnosis. The vast majority of participants had genetic testing and the LQTS type variability closely resembled that of the general population. Previous research has found that 30% to 35% have LQT1, 25% to 30% have LQT2, and 5% to 10% have LQT3 (Yan & Kowey, 2011). In the present study, 32% had LQT1, 34% had LQT2, and 6% had LQT3. The remaining 21% of participants did not know their genotype, and 6% had either LQT4, LQT6, LQT1 and 3, or LQT1, 2, and 3.

Information pertaining to LQTS treatment characteristics was also elicited from participants. These questions assessed presence of ICD/pacemaker, medications and side effects, and information pertaining to LQTS-related medical visits. Among the 91 participants, 60% had an implantable device. An ICD is the first line treatment for individuals with LQTS who have had ongoing symptoms despite use of beta blockers, have experienced cardiac arrest, recurrent syncope, and cannot take beta blockers (Brenyo et al., 2012). The majority of participants reported taking a beta blocker. Thirty-two percent reported a history of cardiac arrest, and 70% have had

syncope. Along with treatment history, AED use was also assessed. Only 18 participants reported that they own an AED. Sixty percent reported that AED use had not been discussed with their cardiologist, 4% reported being unsure if AED use is permitted, and 7% reported that AED use is not permitted by their cardiologist. Reasons for this may include limited discussion between patient and physician, associated cost, and effort it takes to obtain an AED. Future research is needed to identify barriers to discussing AED use in medical visits. The device costs between $1,200 and $2,500 and it is not covered by most insurance companies. There are ways to seek an exception from insurance agencies that results in coverage, with a copay of between $50 and $1,500. There are also grants available for medical patients and organizations. Undoubtedly, many individuals cannot afford an AED and may have barriers to engaging in the effort to obtain one. Future research should investigate barriers to discussing and purchasing the device so that appropriate interventions can be designed.

Quality of life and emotional wellness have been shown to be impacted by various chronic medical conditions (American Academy of Family Physicians, 2011; Moss-Morris et al., 2002; Wilson & Kaplan, 2000). Depressive and anxiety disorders of all types have a lifetime prevalence of 11% and 25%, respectively (American Academy of Family Physicians, 2011). These numbers are likely to increase with the occurrence of chronic medical conditions (Scott et al., 2007; Verhaak, Heijmans, Peters, & Rijken 2005). Among the 91 participants in the present study, 48% reported that they had sought help for emotional issues. The top conditions were anxiety and depression. Participants who reported having sought help for emotional issues were also likely to report heart palpitations, which could be a symptom of LQTS and of

anxiety. The LQTS population would benefit from research that investigates factors that predict emotional issues and/or resilience.

In addition to demographic and illness perception characteristics, adherence characteristics were identified. PA recommendations used to determine adherence in the present study were based on the 36th Bethesda Conference Guidelines for LQTS (Zipes et al., 2005). These guidelines recommend all individuals with electrocardiographically manifested LQTS, whether symptomatic and asymptomatic, are advised to avoid participation in most competitive sports (Zipes et al., 2005) and various PAs of high, moderate, and low intensities (Maron et al., 2004). Specific activities were listed as either not advised, probably permitted, and PAs that require individual clinical assessment (Maron et al., 2004; Appendix B).

Participants in the present study reported participation in each PA identified by the guidelines. The top PAs endorsed, defined as more than 40% participation, were brisk walking, swimming, hiking, treadmill, biking, jogging, bowling, and weightlifting. Participation in lifestyle modifications was also assessed. These reports were compared to participants' reports of their cardiologists' recommendations to determine adherence. The findings indicated that 50% or more of participants had not discussed 18 of 28 PAs on the Bethesda Guidelines list with their cardiologists. Among these 18 activities that were not discussed, 11 were in the not permitted category. Of the PAs and lifestyle modifications that were discussed, 34% of participants endorsed nonadherence. Specifically, nonadherence was found for the following PAs: basketball, soccer, tennis, baseball, swimming, treadmill, hiking, bowling, golf, and snorkeling. Findings indicated that basketball was restricted by participants'

cardiologists more than any other PA on the list (not recommended by 25%). Along with basketball, there was more participation in swimming than any other not permitted activity in the Bethesda Guidelines. Nearly half (49.4%) of participants reported that swimming was permitted by their cardiologists, and 65% of participants endorsed participation in swimming within 12 months prior to the study. Similarly, weightlifting is also in the not permitted category of the Bethesda Guidelines and was a popular activity among participants (41.6% participation). Equal percentages of participants reported that this activity was permitted or not discussed (37.8%). However, PAs listed by the Bethesda Guidelines are not defined by the Bethesda Guidelines and were not defined for participants in this study. Participants could have endorsed participation in low intensity activity (e.g., wading in water and light weightlifting) to high intensity activity (e.g., swimming laps and heavy weightlifting). In addition to identifying PA characteristics, participants also endorsed adherence/nonadherence to other lifestyle recommendations. Consumption of sodium-rich foods and drinking caffeine had the highest nonadherence rates (11% and 10%, respectively).

Per the PA guidelines from 2005, participation in sports with a danger of bodily collision for athletes with an implantable device was also strongly discouraged (Zipes et al., 2005). Among the 91 participants, 60% had an implantable device. Sports that may involve greater risk of bodily collision include basketball, soccer, skiing, ice hockey, motorcycling, horseback riding, and skating. Of these, the highest percentages of participation by adults with an implantable device were soccer 86%, skiing 83%, horseback riding 71%, and basketball 55%. The remaining PAs with risk of bodily

collision had less than 30% participation by adults with an implantable device. There are various potential explanations for adherence and nonadherence.

Sixty-six percent of participants endorsed adherence to medical recommendations. This may have been due to various characteristics of the sample, including participants' value placed on health, the physician/patient relationship, and high ratings of illness coherence. Adherence rates may have been high due in part to participants' value placed on overall health and LQTS treatment. Participants had lower BMIs in comparison to women in 2009 (National Health and Nutrition Examination Survey), endorsed high importance placed on PA, and many reported a moderate to significant change in PA participation after being diagnosed with LQTS. Those who endorsed a change in PA participation stated that LQTS-related factors (e.g., ICD and history of cardiac arrest) contributed to their decisions to change. These findings suggest that participants may value their overall health and LQTS treatment.

Along with value placed on LQTS treatment, the quality of the physician/patient relationship has been shown in various medical populations to predict adherence (Chen et al., 2011; Goldring et. al., 2002; Moine et al., 2007; Orom et al., 2013; Schneider et al., 2004; Steinhauser et al., 2010). For the present study, all measures of the physician/patient relationship, with the exception of agreement on treatment goal, were positively skewed. Participants largely endorsed a positive appraisal of their cardiologists. A notable characteristic of the current sample is that over half of participants reported that they have changed their cardiologist. Chi-square tests determined that change in physician was not significantly related to whether a participant endorsed emotional issues or symptom history. Participants did report that open

communication with their physician was one of the most important factors to keep them safe and healthy. One possible explanation for the high ratings of the physician/patient relationship is that participants in the sample changed cardiologists until they were satisfied with their care. Another possibility for the skewed ratings was that the use of self-report may have elicited a biased response. Along with the impact of the physician/patient relationship on adherence rates, participants high ratings of understanding LQTS may have also contributed.

Illness coherence, or understanding one's medical condition, has been shown to predict medical adherence (Chen et al., 2011; Moss-Morris et al., 2001). Additionally, positive ratings of physician communication have been shown to increase illness coherence (Shipman, 2010). A qualitative study that evaluated common barriers to medical adherence among adults with LQTS found that perceived uncertainty and vagueness of information communicated by their cardiologists was likely to increase patients' risk for engaging in PAs that could have led to a cardiac event (Steinhauser, 2010). Similarly, participants in the present study reported that awareness of LQTS information and frequent communication with their physician were the most important factors to keep them safe and healthy. High ratings of illness coherence were also highly associated with all measures of the physician/patient relationship, excluding agreement on treatment goal. This finding may explain why illness coherence ratings were positive overall with little variability. Furthermore, illness coherence may include knowledge of one's genotype. Research has shown that not knowing which type of LQTS one has may impact treatment adherence (Steinhauser, 2010). For the present study, 74 of the 91 participants reported knowing their LQTS type. Along with factors that may have

influenced adherence, other sample characteristics may have contributed to nonadherence in the current study, including perception of treatment and personal control over LQTS.

Explanations for nonadherence. Overall, 34% of participants in the present study endorsed nonadherence to their cardiologists' recommendations. Prior research has indicated that medical nonadherence is influenced by several factors, including beliefs about treatment and personal control of the medical condition (Goldring et al., 2002; Janz & Becker, 1984; Ronis, 1992), symptom history (Goldring et al., 2002), psychopathology (Watford et al., 2007; Ciechanowski, Katon, & Russo, 2000), and cultural characteristics (Turner et al., 2003).

Studies have indicated that health-related beliefs explain up to 40% of the variance in health protective and maintenance behaviors including adherence (Goldring et al., 2002; Janz & Becker, 1984; Ronis, 1992). This was supported by the present study's findings. A logistic regression demonstrated that the combination of beliefs about the controllability of LQTS by medical treatment and personal behaviors predicted nonadherence. As personal control and treatment control decreased, the likelihood of adherence decreased. This finding is consistent with prior research in various medical populations (Chen et al., 2009; Chen et al., 2011; Moss-Morris et al., 2002; and Ross, Walker, & MacLeod, 2004), including a qualitative study evaluating barriers to treatment among adults with LQTS (Steinhauser, 2010). Furthermore, participants who reported a sense of self-efficacy and belief that their behaviors could control LQTS also endorsed higher rates of illness coherence. There was not a significant relationship between having a clear understanding of LQTS and belief that medical treatment will control the illness. There are possible explanations for this outcome that require further

investigation. Overall, the participants reported positive relationships with their cardiologists. Previous studies have found that the quality of the physician/patient relationship, treatment control, personal control, and illness coherence each influence medical adherence (Chen et al., 2011; Moss-Morris et al., 2001). Research has not been conducted to determine if the quality of the physician/patient relationship mediates or moderates the relationship between treatment control and illness coherence.

Along with perceptions of treatment control and personal control over LQTS, additional factors may have accounted for nonadherence. Research shows that symptom experience is related to nonadherence (Goldring et al., 2002), although findings have been inconsistent (Carney et al., 2010; Haynes & Sackett, 1979). The authors of one study suggested that adherence may predict symptom occurrence, rather than symptoms as a predictor of adherence (Carney et al., 2010). Results from the present do not support this argument, as 77% of the sample was symptomatic and 34% were non-adherent to medical recommendations. Furthermore, a chi-square analysis did not demonstrate a significant relationship between symptom experience and adherence. Various hypotheses may explain this outcome. The impact of symptom experience may vary depending on frequency of symptoms and type of symptoms. Seventy-one percent of the sample experienced symptoms never, one to three times, or yearly. The remaining 29% experienced symptoms weekly to monthly. In addition, the sample in the present study reported low variability with regard to the quality of the physician/patient relationship and largely reported positive relationships. If the physician/patient relationship mediates the association between symptom experience and adherence, this could also explain the

current findings. However, this connection between variables has not been researched and would be useful to evaluate in future adherence studies.

Psychopathology, including depression, anxiety, and personality disorders has been found to impact adherence rates (Ciechanowski et al., 2000; Watford et al., 2007;). According to the self-regulatory model, the combination of cognitive and emotional representations of one's illness often lead to emotion-based coping behaviors in response to perceived illness threats (Leventhal et al., 2001). Among the 91 participants in the present study, 48% reported seeking help for emotional issues. Current findings indicated that participants' more threatening views of LQTS were associated with a negative emotional representation. Negative beliefs about LQTS consequences, symptom unpredictability, and low treatment control were each correlated with a negative emotional representation. The emotional impact of LQTS on some participants may have affected nonadherence to treatment; further investigation is needed. Cultural factors have also been shown to predict medical adherence.

Cultural characteristics, including race, age, gender, socioeconomic status, and race concordance between physician and patient have been found to be related to medical adherence (Cooper et al., 2003; Kaplan et al.,1995; Schneider et al., 2004; Turner et al., 2003). There was not sufficient variability in the data to detect associations with crosstabulaitons and chi-square analyses between adherence and gender, socioeconomic status, and race. This could be due to cultural bias associated with use of online support for medical conditions (Fox, 2011) and possibly low cultural variability among the LQTS population (Brenyo et al., 2012; Schwartz et al., 2016). There may also have been variability among physician race that could have impacted nonadherence (Cooper et al.,

2003). The LQTS population would likely benefit from future research that includes greater cultural variability among patients and physicians. Future investigation regarding nonadherence among individuals with LQTS should include the new PA guidelines (Ackerman et al., 2015).

Updated PA guidelines. The present study was designed based on the Bethesda Guidelines published in 2005 (Zipes et al., 2005). These guidelines were established despite the observation that PA has been found to be a potential trigger for LQTS, particularly LQT1 (Priori et al., 2002; Schwartz et al., 2010). Accumulating data suggested that the guidelines may have been too stringent (Johnson & Ackerman, 2014), as they encompassed all channelopathies without taking into account individual likelihood of risk (Ackerman et al., 2015). Consequently, there has been great variability in physicians' ability to diagnose, risk stratify, and treat individuals with LQTS (Taggart et al., 2007). These findings led to the development of new PA recommendations for various channelopathies, including LQTS (Ackerman et al., 2015).

The new PA guidelines recommend that treatment programs be highly individualized and developed by physicians who specialize in treating cardiac channelopathies (Ackerman et al., 2015). Individuals with LQTS are advised to be treated by a heart rhythm specialist or genetic cardiologist with expertise in LQTS (Ackerman et al., 2015). The guidelines recommend that treatment decisions be made based on the severity of the phenotype and not influenced by the patient's athletic status (Ackerman et al., 2015). For example, patients should not be given an ICD so that they can continue participating in competitive sports. The new recommendations also state that PA participation should be restricted for 3 months for an athlete who experiences a

cardiac event and is suspected of having LQTS or another channelopathy (Ackerman et al., 2015). Once 3 months has passed and the individual has had a comprehensive evaluation, his or her family has been well informed, and a treatment regimen has been established, the patient may be permitted to return to PA participation (Ackerman et al., 2015). For an individual who is symptomatic and/or has EKG-manifested LQTS, competitive PA participation may be continued if the athlete has been asymptomatic for 3 months (Ackerman et al., 2015).

When an individual has a LQTS genetic mutation, but does not have an elongated QT interval on an EKG, and is asymptomatic, this is referred to as concealed LQTS (Aziz et al., 2011). All individuals with LQTS, including athletes with concealed LQTS, should be advised to avoid QT-prolonging substances, dehydration, and exhaustion/heat stroke (Ackerman et al., 2015). They should be encouraged to maintain electrolyte hydration, own an AED, and establish an emergency plan with schools and coaches (Ackerman et al., 2015).

In addition to these guidelines, it is recommended that individuals with LQTS who have an ICD adhere to the Task Force 9 guidelines (Zipes, Link, Ackerman, & Kovacs, 2015). Based on data obtained from the North American ICD Sports Registry, athletes with an ICD may continue PA participation due to the overall low associated mortality, low likelihood that the ICD could be damaged, and low likelihood that inappropriate shocks may occur (Lampert et al., 2013). However, these factors may vary depending on the sport (Zipes et al., 2015). As a result, PA recommendations should be based on evidence for the benefit and risk of the specific sport in which a patient wishes to participate (Zipes et al., 2015). Furthermore, the patient should be counseled on the

likelihood of inappropriate shocks and potential risk for device-related trauma (Zipes et al., 2015). These new guidelines may facilitate tailored treatment recommendations for individuals with LQTS.

Two hypotheses were evaluated with forward multiple regressions to determine if the quality of the physician/patient relationship predicts personal control and treatment control. All predictors except for agreement on treatment goal were removed from the equation of both analyses because assumptions of homoscedasticity and normal distribution were not met. Variables removed were agreement on task with physician, emotional bond, adherence dialogue, overall communication, and overall satisfaction. Results of the analyses indicated that agreement on treatment goal between participant and cardiologist significantly predicted participants' perceptions that both medical treatment and personal behaviors control LQTS. Although these results were significant, agreement on treatment goal accounted for 3% and 4% of the variability of personal and treatment control, respectively. This is supported by prior research.

A qualitative study that investigated barriers to adherence among adults with LQTS found themes based on participants' reports, including perceived low control over LQTS, belief that their physicians were vague, and confusion about treatment recommendations (Steinhauser, 2010). It is possible that low ratings of agreement on treatment goal between patient and physician could be related to uncertainty of treatment goal. Beliefs that the future is uncertain and unpredictable is often accompanied by perceived ineffectiveness at solving problems (Nezu, Nezu, & Lombardo, 2004), thus impacting sense of control. Future research is needed to investigate this relationship.

In addition, when providers use a patient-centered interaction style during medical visits, such as motivational interviewing, that involves eliciting patients' motivating values and conveying empathy and understanding, patients are more likely to be medically adherent (Rollnick, Miller, & Butler, 2008). Findings of the present study may have begun to explain the mechanisms of this relationship. Changing perceptions of treatment and personal control may explain how patient-centered communication and motivational interviewing are effective.

In addition to existing research that supports these findings, many factors also account for the remaining 96% of variability in personal and treatment control, including beliefs about LQTS consequences, physician/patient relationship factors, LQTS and family history, and presence of anxiety. Beliefs associated with LQTS are related to perception of control over LQTS. Participants who believed that LQTS is a serious condition with major interpersonal, financial, and other life consequences also believed that their medical treatment provided little control over LQTS. Beliefs that LQTS resulted in serious life consequences were significantly associated with beliefs that symptoms are unpredictable. Therefore, perceptions of high consequences and unpredictability may have predicted beliefs about control.

The quality of the physician/patient relationship may also contribute to adults' beliefs about control over LQTS. Participants rated their physicians highly on adherence dialogue, overall communication, overall satisfaction, emotional bond, and agreement on treatment task. Research has not been conducted to evaluate which type of physician rating is most likely to influence patients' beliefs about control. It is possible that the remaining physician/patient relationship variables also predict personal and treatment

control. Results of the present study demonstrated a significant association between adherence dialogue and personal control. Specifically, participants who reported that physicians gave information about what PAs should be avoided or continued, understood problems participants have had with following the treatment recommendations, and helped to solve these problems also endorsed the perception that their personal behaviors can control their LQTS and they have a clear understanding of LQTS. Consequently, these additional characteristics of the physician/patient relationship may have also predicted participants' beliefs about control.

An additional explanation for the variability in participants' beliefs about personal and treatment control is that some participants' sense of control may be a trait rather than a state. Participants who have comorbid anxiety may have a tendency to perceive future uncertainty, unpredictability, and a low sense of control (Nezu et al., 2004). Participants who tend to have a negative problem-solving orientation are also more likely to have a low sense of control (Nezu e al., 2004). These perceptions and problem-solving style may account for part of the variance in beliefs about treatment control and personal control.

Undoubtedly, the new PA guidelines may facilitate tailored treatment recommendations for individuals with LQTS, which may result in improved communication between patient and physician and agreement on treatment goal. Additional research is needed to understand all factors that predict adults' with LQTS beliefs about personal and treatment control. The impact of cultural factors would be important to evaluate, particularly those that have been found to be related to locus of

control. Previous research has connected beliefs about control to emotion and behavior (Cheng, Cheung, Chio, & Chan, 2013).

Limitations

Limitations of the present study include participant selection, generalizability, and self-report. Recruitment included various Internet social media groups that have high volumes of individuals with medical conditions. The Internet is often used as a recruitment tool because it reaches a majority of the population and provides comfort and anonymity to research participants (Choules, 2007; Chris & Stewart, 2000; Bender, Jimenez-Marroquin, & Jadad, 2011). Internet research increases the likelihood of reaching people of various cultural backgrounds. However, variability in gender and ethnicity did not occur in this study. In addition, Internet recruitment is likely to result in cultural bias toward women and individuals with higher income, greater health literacy, better overall health, and decreased psychological distress (Griffiths et al., 2012). Only six participants reported a $20,000 annual income or less, and most had at least a bachelor's degree. The gender bias in the study is also a limitation. Data from the present study was largely collected from women. Existing research has shown that women tend to be more non-adherent than men (Turner et al., 2003; Lewey et al., 2013; Lee & Khan, 2015). There is not currently an explanation for this finding. Whether the current findings can be applied to men with LQTS is unknown. Furthermore, very few participants were from rural areas, as most reported living in suburban or urban areas. Due to these sample characteristics, the ability to generalize to individuals who are male, of lower socioeconomic status, live in rural areas, and have less education is uncertain. Working toward reducing health disparities among the LQTS population

through inclusive research is essential to providing quality care. Along with limitations in generalizability related to demographic bias, the study is likely to have prompted participation by individuals who seek support from others who have LQTS. Therefore, results may or may not apply to those who do not seek support.

Along with issues in participant selection and generalizability, there were inherent limitations of self-report measures. All variables of interest were assessed with participant self-report. Self-reported adherence may result in overestimation due to social and personal desirability. In addition, research has shown that patient and physician self-reports are not typically highly correlated with objective ratings of the same encounters (Epstein et al., 2005). Despite these limitations, data from the present study provide a starting point for further understanding of the psychosocial aspects of medical adherence among the LQTS population.

Implications for Psychologists

Health psychology research geared toward identifying comorbid psychological and medical conditions that impact individuals' quality of life, medical prognosis, and overall health is growing. Clinical psychologists who work in medical settings can apply knowledge gained through research to prevent medical nonadherence and improve health outcomes. The biopsychosocial model conceptualizes medical conditions and health behaviors, such as nonadherence (Epstein et al., 2005). This model defines adherence as it relates to patient characteristics, physician characteristics, and health system factors (Epstein et al., 2005). A prominent patient characteristic that has been shown to influence nonadherence is health beliefs. Findings from the present study support this relationship and can be explained by the self-regulatory model.

Leventhal et al. (1992) developed the self-regulatory model to better understand the connection between health behaviors and beliefs. The model suggests that medical patients strive to maintain a state of normalcy by creating an illness identity, implementing strategies to cope with their illness beliefs and emotional impact, and evaluating the effectiveness of the chosen coping strategy. Illness identity is comprised of beliefs related to illness timeline, changeability, consequences, treatment control, personal control, understanding, and emotional representation. Illness-related behaviors, such as nonadherence, have been shown to be specifically influenced by beliefs about personal control, treatment control, and illness coherence (Moss-Morris et al., 2002).

The present study found that perceptions of personal and treatment control predict adherence and nonadherence in adults with LQTS. Furthermore, agreement between patient and cardiologist on treatment goal significantly predicted higher ratings of personal control and treatment control. Based on this information, clinical psychologists who work in medical settings and encounter individuals and families with LQTS can intervene in two critical components of the biopsychosocial model that impact nonadherence. These components are patient characteristics (i.e., health beliefs and emotional impact) and physician characteristics (i.e., facilitate agreement on treatment goal).

The first method of intervention can be implemented by behavioral health specialists to target patient characteristics related to nonadherence. Cognitive behavioral therapy is an empirically based theoretical orientation used by clinicians in both mental health and medical settings. This approach is in line with the self-regulatory model, as it guides patients to identify and modify problematic beliefs, behaviors, and emotional

coping that impact their overall health (Beck, 2011). Use of cognitive-behavioral therapy in medical settings has been shown to decrease adherence rates (Gonzalez et al., 2010; Mohr et al., 2012) and treat many psychological disorders that also impact adherence (Ciechanowski et al., 2000; Watford et al., 2007).

The second method of intervention can be implemented by psychologists and medical providers to target the physician/patient relationship and agreement on treatment goal. Leventhal et al.(1992) speculated that development of illness beliefs is facilitated by experiences related to the illness, which includes interactions with physicians. Findings of the present study indicated that agreement on treatment goal between participant and cardiologist predicted higher ratings of treatment and personal control. Agreement on treatment goal can be facilitated by patient-centered communication by the provider (Rollnick et al., 2008). Motivational interviewing is a patient-centered style of communicating that strengthens patients' motivation to change to improve health-promoting behaviors (Rollnick, Miller, & Butler, 2008). Providers who use motivational interviewing elicit patients' values and align with adaptive values to increase motivation to adhere to treatment. Motivational interviewing can be utilized by all health care providers, including psychologists, physicians, and other medical staff. Motivational interviewing training for various health care providers are offered nation wide and can also be taught by psychologists with expertise in this area within their clinics. Not only do the present findings inform clinical care, but they also inspire future directions with the LQTS population.

Future Directions

The current findings generate many opportunities for future work with the LQTS population that involve research, intervention development, and advocacy. Results indicate that medical adherence is predicted by individuals' beliefs that their medical treatment controls their LQTS and they have personal control over their LQTS. Investigating variables that mediate or moderate the relationship between illness beliefs and adherence would be beneficial. Studies may include variables that have been previously found to predict nonadherence, including cultural characteristics (race concordance of physician/patient, age, socioeconomic status, and gender), patient characteristics in addition to beliefs (psychopathology, problem-solving orientation, symptom history, and health literacy), physician characteristics (personality, experience in the field, style of communication, and duration of patient/physician relationship), and health system factors (waiting times, courtesy of staff, and access to care).

Along with beliefs about control, the perception that the patient understands his or her medical condition has also been found to predict or be associated with medical adherence among other populations (Moss-Morris et al., 2002; Steinhauser, 2010). Illness coherence was not incorporated in the current regression models due to low variability in the data. Future research should investigate the impact of illness coherence on medical adherence among the LQTS population.

In addition, low illness coherence among adults with LQTS may be related to the previous PA recommendations that were criticized for being too general (Zipes et al., 2005). Research to clarify PA recommendations for the LQTS population is advancing (Johnson & Ackerman, 2014). However, there continues to be an overall lack of

evidence that identifies the true risk an athlete with LQTS faces by continuing to participate in specific competitive sports (Ackerman et al., 2015). Increasing evidence in this area is likely to have a lasting impact on the adherence and the quality of life of individuals with LQTS who value PA.

Along with continuing research on medical adherence, continued evaluation of the efficacy of the PA guidelines for LQTS is important. The new PA guidelines state that treatment programs should be highly individualized and developed by physicians who specialize in treating cardiac channelopathies (Ackerman et al., 2015). Individuals with LQTS are advised to be treated by a heart rhythm specialist or genetic cardiologist with expertise in LQTS (Ackerman et al., 2015). Some adults with LQTS may have difficulty adhering to the recommendation. Individual and health system barriers (low socioeconomic status, barriers to access, low health literacy, etc.) to care by specialists has not been evaluated. Consequently, a large portion of individuals with LQTS may not have access to appropriate care. New research should expand demographics to include individuals from underserved populations. Subsequent intervention can target closing gaps in care that individuals in underserved populations face.

In addition to targeting factors that may contribute to adherence and PA recommendations, studies that investigate the impact of motivational interviewing implemented by the physician and/or behavioral specialist on perceptions of treatment and personal control and adherence would also be beneficial. Furthermore, future work may also involve the development of new clinical interventions that apply evidence obtained about medical adherence to medical patients. For instance, the use of online medical groups is increasing and can be utilized by healthcare organizations to facilitate

patient engagement in health promoting behaviors (Rollman, Herbeck, & Rotondi, 2014). A recent article discussed how accountable care organization clinics can create and evaluate the impact of their own online groups that are moderated by healthcare providers (Rollman et al., 2014).

Overall, psychosocial research of the LQTS population is limited. Although the prevalence of this condition is relatively low, the impact on individuals and families can be significant. Research should continue to identify and explain psychosocial issues and treatment barriers that impact individuals with LQTS. This approach may result in earlier diagnoses and improved self-management of the condition, thus decreasing mortality rates.

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Appendix A
Recruitment Letters

Letter to Website/Group Administrator

Volunteers Wanted – Adults with Long QT Syndrome

A diagnosis of Long QT Syndrome (LQTS) impacts individuals' lives in many ways. Learning about the specific needs that adults with LQTS have is important when providing the best healthcare possible. To help healthcare professionals who work with this population better understand patients' needs, a study is being conducted about physical activities, the physician/patient relationship, and beliefs about having LQTS. This study is led by Stephanie Felgoise, Ph.D., ABPP and Maggie Monk, M.S. at the Philadelphia College of Osteopathic Medicine (PCOM).

May I have your permission to post a short recruitment letter on your (website/group page)? If you would like additional information about the study, please contact lqtstudies@pcom.edu.

Letter to Participants
Volunteers Wanted – Adults with Long QT Syndrome

A diagnosis of Long QT Syndrome (LQTS) impacts individuals' lives in many ways. Learning about the specific needs that adults with LQTS have is important when providing the best healthcare possible. To help healthcare professionals who work with this population better understand patients' needs, a study is being conducted about physical activities, the physician/patient relationship, and beliefs about having LQTS. This study is led by Stephanie Felgoise, Ph.D., ABPP and Maggie Monk, M.S. at the Philadelphia College of Osteopathic Medicine (PCOM). *Each participant will have a 1 in 10 chance of receiving a $10 gift card to either Walmart or Target, that can be used in stores or online.*

Participants must:
- Have been diagnosed with LQTS
- Be at least 18 years old
- Reside and have a doctor for their LQTS in the United States

Participation is ANONYMOUS and the survey takes approximately 20 minutes to complete

To participate in the study, click on the link below.
(link)

If you would like more information about the study, email lqtstudies@pcom.edu.

Appendix B
Adherence

In regard to your LQTS, what do you think is the most important factor to keep you safe and healthy? _____

How often do you participate in physical activities?

Daily
Weekly
Monthly
Bi-Monthly (about once every 2 months)
Bi-Annually (about once every 6 months)
Annually
Never

What are the top 3 physical activities that you participate in the most?
1._____
2._____
3. _____

Are you an active member of a gym? _____ yes _____no

Using the scale below, in the last 12 months, approximately how often have you participated in the following?:

Daily
Weekly
Monthly
Bi-Monthly (once every 2 months)
Bi-Annually (once every 6 months)
Annually
Never

Basketball
Body Building
Ice Hockey
Racquetball/squash
Rock Climbing
Running
Skiing
Soccer
Tennis
Touch Football
Windsurfing

Baseball/softball
Biking
Motorcycling
Jogging
Sailing
Surfing
Swimming
Treadmill
Weightlifting
Hiking
Bowling
Golf
Horseback Riding
Scuba Diving
Skating
Snorkeling
Brisk Walking
Ate foods high in potassium (bananas, spinach, white beans, baked potatoes with skin, apricots, plain yogurt, salmon, avocado, mushrooms)
Ate foods high in sodium
Consumed grapefruit/grapefruit juice

Drank alcohol
If yes, how many days per week do you drink alcohol? _____
How many alcoholic beverages do you consume each day that you drink? _____

Wore a LQTS identification item
Taken an external defibrillator on outings
Consumed Caffeinated Products (Coffee, Tea, Energy Drinks, Chocolate)
Taken recreational drugs or prescriptions not prescribed by your physician
If yes, please name _____

How much water do you drink daily? _____ ounces
How much gatorade do you drink daily? _____ounces

How often do you *remember* to take medication for your LQTS?
_____ Once Daily
_____ Two to Three Times Daily
_____ Other; Please specify _____

Do you have a pacemaker or ICD? _____ yes _____no
If yes, how often do you *remember* to check your pacemaker or ICD? _____

Individuals who report that they participate in an activity that is not permitted by their cardiologist will be asked why they participate in the activity.

Of the top 3 physical activities that you listed that you participate in the most, does your *current* cardiologist recommend that they are Permitted, Not Permitted, or are you Unsure if they are permitted.

Permitted Not Permitted Unsure

Activity #1._____
Activity #2._____
Activity #3. _____

Does your *current* cardiologist recommend that the following activities are Permitted, Not Permitted, or are you Unsure if they are permitted?

	Permitted	Not Permitted	Unsure	Not Discussed
Basketball				
Body Building				
Ice Hockey				
Racquetball/squash				
Rock Climbing				
Running				
Skiing				
Soccer				
Tennis				
Touch Football				
Windsurfing				
Baseball/softball				
Biking				
Motorcycling				
Jogging				
Sailing				
Surfing				
Swimming				
Treadmill				
Weightlifting				
Hiking				
Bowling				
Golf				
Horseback Riding				
Scuba Diving				
Skating				
Snorkeling				
Brisk Walking				
Eating foods high in potassium (bananas, spinach, white beans, baked potatoes with skin, apricots,				

plain yogurt, salmon, avocado, mushrooms)
Eating foods high in sodium
Consuming grapefruit/grapefruit juice
Consuming alcohol
Smoking cigarettes
Wearing a LQTS identification item
Taking an external defibrillator on outings
Consuming Caffeinated Products (Coffee, Tea, Energy Drinks, Chocolate)
Taking recreational drugs or prescriptions that are not prescribed to you by a physician

How much water does your cardiologist recommend that you drink daily? _____ Ounces
_____ Unsure

How much gatorade does your cardiologist recommend that you drink daily?_____ Ounces
_____ Unsure

How often does your cardiologist recommend that you take medication for your LQTS?
_____ Once Daily
_____ Two to Three Times Daily
_____ Other; Please specify _____

If you have a pacemaker or ICD, how often does your cardiologist recommend that you check it?
_____
_____ I don't have a pacemaker or ICD

How often have you participated in your top 3 physical activities since you were diagnosed with LQTS? (So, if you swim monthly, but walk daily, answer with the most frequent activity, i.e., daily).

Daily
Weekly
Monthly
Bi-Monthly (once every 2 months)
Bi-Annually (once every 6 months)
Annually
Never

How often have you participated in the following activities since being diagnosed with LQTS?

Daily
Weekly
Monthly
Bi-Monthly (once every 2 months)
Bi-Annually (once every 6 months)
Annually
Never

Basketball
Body Building
Ice Hockey
Racquetball/squash
Rock Climbing
Running
Skiing
Soccer
Tennis
Touch Football
Windsurfing
Baseball/softball
Biking
Motorcycling
Jogging
Sailing
Surfing
Swimming
Treadmill
Weightlifting
Hiking
Bowling
Golf

Horseback Riding
Scuba Diving
Skating
Snorkeling
Brisk Walking
Eating foods high in potassium (bananas, spinach, white beans, baked potatoes with skin, apricots,
plain yogurt, salmon, avocado, mushrooms)
Eating foods high in sodium
Consuming grapefruit/grapefruit juice
Drinking alcohol
Smoking cigarettes
Wearing a LQTS identification item
Taking an external defibrillator on outings
Consuming Caffeinated Products (Coffee, Tea, Energy Drinks, Chocolate)
Taking recreational drugs or prescriptions that are not prescribed to you by a physician
Drinking Water
Drinking Gatorade

In thinking about your frequency of physical activity before your diagnosis of LQTS compared to after your diagnosis of LQTS, how much do you think your frequency (how often) of physical activity has changed?

1 2 3 4 5
Not at all Very Much

How important is participation in physical activities to you?

Not at all important
Low importance
Slightly important
Neutral
Moderately important
Very important
Extremely important

Does your cardiologist discuss your physical activities with you? _____ yes _____ no

Appendix C
Personal Information Questionnaire

Age: _____

Gender: Male Female

Weight: _____

Height: _____

Which group best describes how you identify yourself?
Native American
Asian (includes Pacific, South, Southeast, and North)
Black
Hispanic (Nonwhite)
White
Middle Eastern
Multiracial

Which group best describes your religious beliefs?
Buddhist
Christian
Hindu
Jewish
Muslim
Agnostic
Atheist
Other, please list: _____

Which best describes your current employment status? (check all that apply)
Unemployed student
Unemployed
Employed (full-time)
Employed (part-time)
Retired
On disability

Are you in school?
Yes No
If yes, what grade/level?
High School College Graduate-Master's program Graduate-Doctoral program
Other _____
Highest academic degree you have earned:
High school diploma/GED
Associate degree (2 years of college)

Bachelor's degree (4 years of college)
Master's degree (Please specify what discipline: _____)
Doctoral degree (Please specify what discipline: _____)

What is your yearly household income range?
$5,000-$20,000
$21,000-40,000
$41,000-$60,000
$61,000-$80,000
$81,000-$100,000
Above $100,000

Which residential area of the choices below best describes where you live?
Farm
City
Rural
Suburban
Other

If you live in the United States, what region of the country do you live in?
Northeast
South
Midwest
West

At what age were you first diagnosed with LQTS? _____

What was the date of your last scheduled cardiology/LQTS related medical visit?
_____

Have you had genetic testing to confirm your LQTS diagnosis? Yes No

What type of LQTS do you have or does your doctor suspect you have?
LQTS 1 LQTS 2 LQTS 3 LQTS 4 LQTS 5 LQTS 6 LQTS 7
Unidentified gene Other _____

Do you have any other diagnosed medical condition?
Yes No
If Yes, please describe:
_____
_____
_____

Do you think you have any other UNDIAGNOSED medical condition(s)?
Yes No
If Yes, please describe:
_____
_____
_____

If you have ever had symptoms of your LQTS, answer the following questions:
When was your first symptomatic episode (i.e. fainting, sudden cardiac arrest, seizure)?
_____

In what setting did this episode occur?
School Home Sporting Activity Work
Other, please specify: _____

How old were you at the time of your first LQTS event or symptom? _____

Did you go to the Emergency Room after your first LQTS event? Yes No

When was your most recent event? Date: _____ (month) _____(Year)

How frequently do your LQTS symptoms occur?
Never 1-3 times Weekly Monthly Yearly

How many events (FAINTING, SEIZURE, ARREST) in total have you had?
0 1 or 2 3 or4 5 or 6 7 or more

Do you have a pacemaker or implantable cardioverter defibrillator (ICD)?
Pacemaker ICD Both Neither

Do you have an Automated External Defibrillator (AED)?
Yes No

If yes, Please describe when you carry it with you:
_____
_____
_____

What medication(s) do you take for LQTS or other medical conditions?
_____
_____
_____

Do you experience side effects from medications? If so, please specify.
Yes No
If yes, please specify: _____

How often do you see your cardiologist?
Every 3 months Every 4 months Twice a year Yearly Other

Have you changed your cardiologist since being diagnosed with LQTS? _____ yes
_____no

If yes, how many cardiologists have you seen since being diagnosed with LQTS? _____

When did you begin seeing your current cardiologist? _____

What other professionals (non-cardiologists) do you discuss LQTS with? _____

How often do you discuss LQTS with other professionals? _____

Have you sought help from a medical or mental health professional for emotional problems, such as anxiety, depression, or other? If so, please specify.
Yes, please specify_____
No

Who lives in your household? Please check all that apply.
Parents Siblings Roommate(s) Live Alone
Spouse/Partner Children Other _____

How many children, if any, do you have?
None 1 or 2 3 or 4 5+

How many of your children have LQTS? _____

If you have children that have a LQTS diagnosis, are they symptomatic?
Yes No

If you have children that have a LQTS diagnosis, do they see the same cardiologist as you?
Yes No

Do members in your immediate and/or extended family have LQTS? Yes No

Are there members of the family with LQTS who have <u>never</u> had symptoms?
Yes No

Have you participated in other LQTS studies conducted by the Philadelphia College of Osteopathic Medicine?
Yes No Unsure

Appendix D
Twitter Recruitment Post

#LongQTSyndrome Study! Adults Needed to Complete Survey about Physical Activity/Dr. Satisfaction: https://www.surveymonkey.com/s/LQTS-STUDY