

2015

A Qualitative Study of How Children Experience and Live with Long QT Syndrome

Patrick McElwaine

Philadelphia College of Osteopathic Medicine, patrickmce@pcom.edu

Follow this and additional works at: http://digitalcommons.pcom.edu/psychology_dissertations

 Part of the [Cardiology Commons](#), [Child Psychology Commons](#), and the [Health Psychology Commons](#)

Recommended Citation

McElwaine, Patrick, "A Qualitative Study of How Children Experience and Live with Long QT Syndrome" (2015). *PCOM Psychology Dissertations*. Paper 336.

This Dissertation is brought to you for free and open access by the Student Dissertations, Theses and Papers at DigitalCommons@PCOM. It has been accepted for inclusion in PCOM Psychology Dissertations by an authorized administrator of DigitalCommons@PCOM. For more information, please contact library@pcom.edu.

Philadelphia College of Osteopathic Medicine

Department of Psychology

A QUALITATIVE STUDY OF HOW CHILDREN EXPERIENCE AND LIVE
WITH LONG QT SYNDROME

By Patrick McElwaine

Submitted in Partial Fulfillment of the Requirements for the Degree of

Doctor of Psychology

June 2015

**PHILADELPHIA COLLEGE OF OSTEOPATHIC MEDICINE
DEPARTMENT OF PSYCHOLOGY**

Dissertation Approval

This is to certify that the thesis presented to us by PATRICK MCELWAINÉ
on the 7th day of May, 2015, 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.

Committee Members' Signatures:

Stephanie H Felgoise, PhD, ABPP, Chairperson

Susan Panichelli Mindel, PhD

Charmaine Chan, DO

Robert A DiTomasso, PhD, ABPP, Chair, Department of Psychology

Dedication

This dissertation is dedicated to the children who volunteered to participate in this study. It is through their courage, strength, and voice that we gain insight into how a child lives with and experiences long QT syndrome (LQTS). Through their participation in this study, these children are helping other children who are living with LQTS, helping the parents of those with LQTS to better support their children, and providing professionals with insights and information to better provide the best services possible.

Thank you!

Acknowledgements

Completion of this dissertation would not have been possible without the unconditional love and support of numerous individuals in my life. First, I want to express my appreciation and gratitude to my dissertation committee. It is only through the guidance and commitment of Dr. Stephanie Felgoise, Dr. Susan Panichelli Mindel, Dr. Victoria Vetter, and Dr. Charmaine Chan that my dissertation has been successfully completed.

It is very important for me to thank the PCOM faculty for their unbelievable work in strengthening and sharpening my knowledge and skills as a future psychologist, with the main goal of developing the “gold standard” in the field of psychology. I am also extremely thankful and grateful to Dr. Stephanie Felgoise for her vision, time, encouragement, and direction not only in the completion of my dissertation, but also the support she has offered me throughout the doctoral program.

I also want to state my appreciation and gratitude to Elizabeth Waldron and Katherine Corvi, who dedicated their time, energy, and insights as members of my coding team.

There are too many friends and family that have been supportive, encouraging, patient, and loving to name them all in this section, but there are a few I would like to specifically acknowledge. I would like to thank my father, Patrick McElwaine. I know you are looking down on me with a smile today! I would also like to express my appreciation for the guidance and love that my mother has given me throughout my life and throughout my journey in my doctoral program. You are my role model and have instilled in me the resilience, perseverance, and dedication needed to earn my doctorate.

I would like to thank my sister, Melissa McElwaine, and my brother, Steven McElwaine, for their continued support and love. I would also like to express my appreciation to my in-laws, William Anderson, Andrea Anderson, Carrie Bean, and Shawn Bean, for their continued support and encouragement.

I would like to thank my family, Elyse, Morgan, Eva, and my dog Vinnie. I could not have done this without my wife, Elyse McElwaine. You are an amazing, beautiful woman who I feel incredibly lucky to have in my life. I do not have the words to appropriately thank you for the love, support, and sacrifices you have made throughout this journey. To my children, Morgan and Eva: you are the joy and happiness that keep me moving towards being a better man and the best father I can be. Words cannot express how much I love you and how you have helped me throughout my doctoral experience. I love you with all my heart!

Abstract

Long QT syndrome (LQTS) is an inherited and potentially fatal disorder affecting approximately 1 in 2,000 people. Children often experience anxiety and a sense of loss of control as they try to manage the medical, social, and psychological stress that accompanies being diagnosed with LQTS. The present study utilized a qualitative research design to examine how children experience and live with long QT syndrome. Semistructured interviews were conducted with eight children between the ages of 7 to 12 who were diagnosed with LQTS. This period in a child's life is very important for developing social skills and self-esteem as peers and teachers become more important to the child. Themes that emerged involved treatment, relationships, and social connectedness. Children communicated aspects of treatment that were important to them, such as the doctor-patient relationship, stress test, medication, going to the hospital, and lifestyle restrictions. Parental and peer relationships were also explored with regard to how much information should be known by the child, perceptions of worry, confiding in friends or keeping it private, and values of importance in a peer relationship. The children voiced fears about not being accepted and being treated differently and more specific fears, including people knowing about the diagnosis, having to answer questions about the heart monitor, and experiencing feelings of sadness and loneliness. The "Five Pillars of Adaptation for Long QT Syndrome" born from this research are developmental level and self-esteem, peer and social relationships, parental support, social problem-solving, and treatment and resources. They provide the fundamental elements through which we can learn how a child lives with long QT syndrome.

Table of Contents

Dedication	iii
Acknowledgements	iv
Abstract	vi
List of Tables	x
Chapter 1	1
Introduction	1
Statement of the problem.	1
Purpose of the study.....	2
Definitions.....	3
Literature review.....	5
Resiliency, protective, and risk factors.	11
Summary of the literature review.	14
Research question.	15
Chapter 2.....	16
Method.....	16
Design.	16
Data analysis.	16
Participants.....	18

Measures.	22
Procedure.	23
Chapter 3.....	25
Results.	25
Interviews.....	25
Initial impressions.....	26
Living with LQTS: A child’s perspective.....	26
Study population.	27
Chapter 4.....	34
Discussion	34
Developmental stage.....	34
Cognitive behavioral strategy of coping	35
Magical power.	35
Themes.....	36
Theoretical analysis of findings.....	43
The Five Pillars of Adaptation for Long QT Syndrome.....	44
Limitations.....	54
Replication of study.....	56
Future directions.	56
References.....	58

Appendix A.....	70
Personal Information Questionnaire.....	70
Appendix B.....	78
Interview Questions.....	78

List of Tables

Table 1. PedsQL Summary of Participant Demographics and LQTS Information21

Table 2. Child Self-Report PedsQL 4.0 Generic Core Scale and PedsQL 3.0
Cardiac Module Scores28

Table 3. Parent-Proxy Report PedsQL 4.0 Generic Core Scale and PedsQL 3.0
Cardiac Module Scores30

Table 4. PedsQL 4.0 Family Impact Module.....32

Chapter 1

Introduction

Statement of the problem.

Long QT syndrome (LQTS) is an inherited and potentially fatal disorder affecting approximately 1 in 2,000 people (Giudicessi & Ackerman, 2013). Named for its display on an electrocardiogram (ECG) as a prolongation of the QT interval (Giuffre, Gupta, Crawford, & Leung, 2008), LQTS may cause sudden and unanticipated loss of consciousness and/or death (Vincent, 2005). Children diagnosed with LQTS often experience symptoms of anxiety, such as worry and a sense of loss of control (Andersen, Øyen, Bjorvatn, & Gjengedal, 2008). Farnsworth, Fosyth, Haglund, and Ackerman (2006) stated that parents have concerns about finding appropriate psychological treatment for their children diagnosed with LQTS who report symptoms of anxiety. Increased research in the area of psychosocial consequences of LQTS is needed to better inform parents and physicians on how to support these children; more specifically, there is no current research that determines possible protective and resiliency factors for managing medical, social, and psychological stress of children diagnosed with LQTS.

Resiliency has been described in the literature as the ability to “bounce back successfully despite exposure to severe risks” (Bernard, 1993, p. 44). Resilience literature describes resilient children as possessing factors of social competence, problem-solving skills, autonomy, and a sense of purpose and future (Bernard). Children were able to overcome significant stressors, such as poverty, abuse, and physical handicap, and develop into healthy and well-balanced adults (Bernard). Traits of

hardiness and self-enhancement and positive emotions such as laughter and happiness help to foster resilience (Bonanno, 2004). However, no research specifically addresses resiliency factors of children with LQTS.

The U.S. Department of Health and Human Services Administration for Children and Families (2012) defines protective factors as “conditions that, when present in families and communities, increase the health and well-being of children and families” (p. 4). The presence of protective factors is considered to be a positive predictor of who will exhibit resiliency (Earvolino-Ramirez, 2007). Protective factors that aid children in overcoming adversity include positive family circumstances and family dynamics (Engle, Castle & Menon, 1996). Protective factors of children also include possessing average to above average intelligence, strong motivation and internal locus of control, positive self-esteem, supportive siblings, and social support from peers and adults (Werner, 2000). No current literature specifically addresses protective factors for children diagnosed with LQTS. Knowledge, insight, and understanding of how children cope with a life-threatening illness and identifying protective and resiliency factors for coping, adapting, and/or accepting their condition would aid parents of children with LQTS in helping their children adjust to the diagnosis of LQTS.

Purpose of the study.

The purpose of this study was to understand how children aged 5 to 12 years experience living with LQTS. Learning how children with LQTS cope with and manage psychosocial stressors would provide parents, families, other loved ones, schools, treatment providers, and society at large a framework for better understanding of and insight into the psychosocial aspects of an LQTS diagnosis.

Definitions.

Automated external defibrillator (AED): a portable apparatus used to restart a heart that has stopped. It is programmed to analyze cardiac rhythms automatically and indicate to a health professional when to deliver a defibrillating shock after the health professional has determined that no one is in contact with the patient (Mosby's Medical Dictionary, 2009).

Cardiologist: a physician who specializes in the diagnosis and treatment of disorders of the heart (Mosby's Medical Dictionary, 2009).

Chronic illness: any disorder that persists over a long period and affects physical, emotional, intellectual, vocational, social, and spiritual functioning (Mosby's Medical Dictionary, 2009).

Diagnosis: identification of a disease or condition by a scientific evaluation of physical signs, symptoms, history, laboratory test results and procedures. The art of naming a disease or condition (Mosby's Medical Dictionary, 2009).

Epidemiology: the science concerned with the study of the factors determining and influencing the frequency and distribution of disease, injury, and other health-related events and their causes in a defined human population. Also, the sum of knowledge gained in such a study (Dorland's Medical Dictionary for Health Consumers, 2007).

Etiology: the study of all factors that may be involved in the development of a disease, including the susceptibility of the patient, the nature of the disease agent, and the way in which the patient's body is invaded by the agent (Mosby's Medical Dictionary, 2009).

Holter monitor: trademark for a device for making prolonged electrocardiograph recordings (usually 24 hours) on a portable tape recorder while the patient conducts daily activities. The patient also may keep an active diary for the purpose of comparing daily events with electrocardiographic tracings (Mosby's Medical Dictionary, 2009).

Implantable cardioverter-defibrillator (ICD): a surgically implanted electronic device that automatically terminates lethal ventricular arrhythmias by delivering low-energy shocks to the heart, restoring proper rhythm when the heart begins beating rapidly or erratically (Mosby's Medical Dictionary, 2009).

Long QT syndrome (LQTS): an inherited cardiac disorder in which a defect in potassium ion channels interferes with the transmission of electrical signals to the heart muscle, producing a prolonged Q-T interval on an electrocardiogram and sometimes causing cardiac arrhythmias (American Heritage Medical Dictionary, 2007).

Pacemaker: an electrical device that has electrodes attached to the heart to electrically stimulate the heart to beat normally (Gale Encyclopedia of Medicine, 2008).

Quality of life: the degree of satisfaction an individual has regarding a particular style of life. Although assessment tools are available to evaluate physical and social dimensions, an individual's general sense of well-being or satisfaction with the attributes of life is more difficult to evaluate (Miller-Keane Encyclopedia and Dictionary of Medicine, Nursing, and Allied Health, 2003).

Sudden cardiac arrest (SCA): an abrupt, complete loss of heart function that results in loss of blood circulation within the body. An episode of sudden cardiac arrest may be preceded by arrhythmias, including ventricular tachycardia or fibrillation. It is

not caused by the blockage of coronary arteries. Sudden cardiac arrest is reversible in most patients if it is treated within minutes (Mosby's Medical Dictionary, 2009).

Syncope: a brief lapse in consciousness caused by transient cerebral hypoxia. It may be caused by many different factors, including emotional stress, vagal stimulation, vascular pooling in the legs, diaphoresis, and a sudden change in environmental temperature or body position. Also called fainting (Mosby's Medical Dictionary, 2009).

Torsade de pointes: a type of ventricular tachycardia with a spiral-like appearance ("twisting of the points") and complexes on an electrocardiogram that at first looks positive and then negative. It is precipitated by a long Q-T interval, which often is induced by drugs (quinidine, procainamide, or disopyramide), but which may be the result of hypokalemia, hypomagnesemia, or profound bradycardia. The first line of treatment is intravenous magnesium sulfate, as well as defibrillation if the patient's condition is unstable (Mosby's Medical Dictionary, 2009).

Literature review.

History of LQTS.

The first report of long QT syndrome was believed to be by Friedrich Ludwig Meissner in 1856, who described the sudden death of a deaf girl who was reprimanded in school, along with the reports of her two brothers who died suddenly, on separate occasions, after a violent outburst (QTsyndrome.ch, 2007). The first official report of LQTS occurred in 1957 in Norway by Anton Jervelle and Fred Lange-Nielsen, who identified a Norwegian family in which three of the four children were deaf, died suddenly, and had QT prolongation on an ECG (Vincent). In 1963, Romano and colleagues described sudden death among individuals with normal hearing who had QT

prolongation (Vincent, 2005). In the 1960s, LQTS was called the Jervell and Lange-Nielsen syndrome and Romano-Ward syndrome (Mauriello, Johnson, & Ackerman, 2011). The International LQTS Registry was created in 1979 as a long-term project to gain better insight into the research and treatment of long QT syndrome (Moss & Schwartz, 2005). The registry is a comprehensive listing of worldwide LQTS cases that includes patient demographic information, symptoms, and treatment and is a source of comparison data for physicians and individuals diagnosed with LQTS and their families (Lazzara, 2008). The registry has made possible the identification of varying types of LQTS, specific risk factors, clinical course, and lifestyle adjustments necessary for living with LQTS (Moss, Schwartz, Crampton, Locati, & Carleen, 1985).

Epidemiology and etiology of LQTS.

Long QT syndrome is a life-threatening cardiac channelopathy. The number of affected individuals has increased since 2006, when it affected 1 in 2,500 people (Farnsworth et al., 2006). LQTS may cause unexpected loss of consciousness and sudden death (Vincent, 2005). LQTS is most commonly hereditary; however, spontaneous mutations can occur (Farnsworth et al.).

Types of LQTS.

There are currently 13 known types of inherited or acquired LQTS (Knoche, Orland, January, & Maginot, 2012), but LQTS 1, LQTS 2, and LQTS 3 are the most common types (National Heart Lung and Blood Institute, 2011). Each of these three types has different triggers. LQTS 1 is triggered by exercise or emotional stress, making the heart beat fast and initiating abnormal heart rhythms (Moss, 2003). Abnormal heart rhythms due to a surprise or other extreme emotions are associated with LQTS 2 (Moss).

Individuals with LQTS 2 are advised to avoid loud or jolting noises from telephones, clocks, and other devices that may initiate a startled reaction (National Heart Lung and Blood Institute). A slowed heart rate during sleep or rest can trigger the abnormal heart rhythm in LQTS 3 (Moss). LQTS often presents in childhood and manifests with unexplained fainting that may result in sudden cardiac death; 10% of individuals experience death as their first symptom (Mayo Clinic, 2013).

Gender effects.

The risk of a cardiac event decreases with age; however, females are more likely than males to experience such an event (Zareba et al., 2003). LQTS is more often found in women, and women are more likely to experience syncope or sudden death after childbirth or during menstruation (Mayo Clinic, 2013). Zareba et al. (2003) conducted a study through the International LQTS Registry, exploring age and gender differences of 566 children under the age of 15 with the diagnosis of LQTS. They found that the risk for a cardiac event for children under the age of 15 was highest in males (56%) than in females who have LQT1. They also found similar cardiac risk levels in male and female children with LQT2; however, they found a low risk of cardiac events among children with LQT3 (Zareba et al.).

Risk factors for LQTS.

For children diagnosed with LQTS who experience recurrent cardiac events, the most prevalent risk factors are QTc interval > 500 msec, history of an experienced syncope, and aborted cardiac arrest (Wedekind et al., 2009). Research has also shown that despite beta-blocker therapy and QTc duration, after the occurrence of the first

cardiac event, risk for recurrent syncope is increased and can be a high predictor of fatal outcome for children and adolescents with LQTS (Liu et al., 2011).

Diagnosis of long QT syndrome.

The first LQTS diagnostic criteria were presented in 1985 and include both major and minor criteria (Schwartz, Moss, Vincent, & Crampton, 1993). Major criteria for diagnosis are prolonged QT interval ($QTc > 440$ msec), stress-induced syncope, and family members with LQTS; minor criteria for diagnosis are congenital deafness, episodes of T-wave alternans, low heart rate in children, and an abnormal ventricular repolarization (Schwartz et al., 1993). For an LQTS diagnosis to be made, individuals need to meet two major criteria or one major criterion and two minor criteria (Schwartz et al.). In 1993, diagnostic criteria for LQTS were updated to reflect new knowledge (Moss, 2003). The new criteria assign points for various ECG, clinical, and family history findings (Schwartz et al.). Scores of 1 and below indicate low probability, 2 to 3 points denote intermediate probability, and scores of 4 points and above indicate a high probability of LQTS (Schwartz et al.). Currently, cardiologists specializing in heart disease use an ECG, medical history, physical examination, and genetic testing to properly diagnose LQTS (National Heart Lung and Blood Institute, 2011). The diagnosis of LQTS is determined by a prolonged QTc interval or the presence of a genetic mutation of LQTS (Jackson, Huisman, Sanatani, & Arbour, 2011). LQTS has also been diagnosed by examining the family history of LQTS in conjunction with an abnormal presentation on ECG during initial assessment (Etheridge et al., 2007). The initial diagnosis of LQTS is made after the first episode of syncope, and a small percentage of diagnoses are made after sudden death or resuscitated sudden death (Etheridge et al.).

Long QT syndrome may be present, yet undiagnosed, in some children and adults (Farnsworth et al., 2006). More than half of the annual 8,000 unexplained sudden deaths in children were related to LQTS, and children with undiagnosed LQTS have a significantly increased risk of fatality (Johnson & Hanan, 2001). Johnson and Hanan also stated that 60% of these children had a family history of LQTS and experienced symptoms such as dizziness and seizures. Historically, there has been a frequent delay in diagnosis, and in some cases patients with syncope are misdiagnosed with epilepsy (Schwartz et al., 1993). Further, a misdiagnosis of other disorders as LQTS can have significant repercussions, such as lifestyle changes, emotional concerns, and invasive medical procedures that may result in death (Taggart et al.). Physicians should continually question the diagnosis, reassessing individuals with every new piece of data, in order to provide the most accurate diagnosis and in turn the best medical treatment (Vetter, 2007).

Treatment of long QT syndrome.

Beta-blocker administration is an effective form of treatment for reducing risk of cardiac-related LQTS events in children (Wedekind et al., 2009). All individuals diagnosed with LQTS, with very few medical exceptions, should be treated with beta blockers (Crotti, Celano, Dagradi, & Schwartz, 2008). Research has shown that beta-blocker therapy has been most effective in reducing cardiac events among individuals with LQTS 1 and 2 (Moss, 2003). Beta-blocker therapy to “suppress the adrenergic-mediated triggers” (Liu et al., 2011, p. 949) has been observed to reduce lethality in high-risk LQTS children. Research has shown that compliance with beta-blocker treatment of LQTS and avoiding medications that are QT-prolonging drugs decrease the risk of

fatality due to LQTS (Jackson et al., 2011). Drugs that prolong the QT interval are mostly antibiotics and antidepressants; use of QT-prolonging drugs increases the risk of sudden death in individuals with LQTS (Jackson et al). Use of QT-prolonging drugs is a significant risk factor for individuals born with LQTS and has been shown to cause acquired LQTS (Johnson & Hanan, 2001). Many medications in the classes of antiarrhythmics, psychotropics, antihistamines, antimicrobials, and antifungals may induce QT prolongation (Johnson & Hanan).

The most effective treatment for high-risk LQTS individuals is a multimodal approach with an implanted cardioverter defibrillator and beta blockers, along with avoidance of QT-prolonging drugs and appropriate lifestyle changes (Moss, 2003). There is an extensive list of both prescription and over the counter medications that must be avoided because they trigger abnormal heart rhythms, QT prolongation, or adrenaline-like effects (CredibleMeds, 2013). These medications include, but are not limited to, medications to treat allergies, high cholesterol, high blood pressure, depression, and arrhythmias (National Heart Lung and Blood Institute, 2011). Foods and drinks that contain caffeine must also be avoided because they may inhibit the effectiveness of LQTS medication or increase heart rate (Irish Heart Foundation, 2013). In addition to dietary and medication restrictions, activities of daily living (ADLs), such as athletics, work, and exercise, and loud and startling noises need to be restricted or monitored for some (Anderson et al., 2008). Research has consistently shown that an implantable cardioverter-defibrillator is superior to all other forms of therapy for those at risk of heart-related sudden death (Sears & Conti, 2002). The highest risk of sudden death is

within 2 years of the first cardiac episode; the lifetime risk of sudden death is between 15% and 70% (Jackson et al., 2011).

Summary of LQTS research.

Since the first believed report of LQTS in 1856 by Friedrich Ludwig Meissner, the awareness of and information about LQTS has grown exponentially (QTsyndrome.ch, 2007). Through the years, developments have included improvements in diagnosis, medications, lifestyle changes, and treatment options. LQTS is a potentially lethal cardiac heart condition that is overdiagnosed, underdiagnosed, and misdiagnosed and can cause fainting, seizures, and sudden death (Mauriello et al., 2011). Continued emphasis on education and awareness concerning the lethality of LQTS is warranted in order to obtain more reliable and accurate diagnoses and the best possible medical treatment (Vetter, 2007). Despite the expanding literature on the treatment and diagnosis of LQTS, research on the psychosocial factors related to this sudden death cardiac disorder is still lacking (Farnsworth et al., 2006).

Resiliency, protective, and risk factors.

There is a need to identify at-risk children and a need for program development within communities and schools to promote protective factors and resiliency to produce more successful children (Kumpfer, 1999). Because there is a substantial amount of evidence-based research available regarding protective factors and resiliency, development of a best practice guideline focused on harm reduction and promotion of protective and positive factors would aid in raising better-adapted children (Resnick, 2000). The “future success of our country depends on increasing our technology and

interest in building better children . . . resilience and child development is critical to the prevention field and our nation's prosperity and well-being" (Kumpfer, 1999 p. 214).

Resiliency research has increased, but there is not yet consensus for an operational definition (Herrman et al., 2011). Despite the lack of consensus, most researchers agree that there is an interplay of protective factors and risk factors within biological, psychological, and social systems that contribute to an individual being resilient (Herrman et al.).

Resiliency.

Over the years, *resilience* has replaced *invulnerable* when describing how individuals can persevere in extremely difficult situations (Earvolino-Ramirez, 2007). Adversity, which is the foremost antecedent, contains challenges, changes, and disruptions through which individuals can develop and utilize the resiliency they possess (Earvolino-Ramirez). The salient internal attributes of resiliency are self-efficacy, hope, and coping; these can be developed at any time in life and are greatly influenced by protective and risk factors (Gillespie, Chaboyer, & Wallis, 2007).

An individual who exhibits resilience is able to realistically assess and analyze the situation and holds realistic expectations as to the outcome (Gillespie et al., 2007). Resilient individuals often have qualities such as a good sense of humor, positive relationships and social supports, self-determination, positive self-esteem, and flexibility in adapting to change (Earvolino-Ramirez, 2007). The principal emerging outcomes of resilience are development and enhancement of personal control, psychological adjustment, and personal growth (Gillespie et al.). Salient effects of resiliency are the abilities to cope and positively adapt to new situations (Earvolino-Ramirez).

Protective factors and risk factors.

Of great concern for children diagnosed with LQTS or congenital cardiac disease is what pediatric healthcare professionals have been calling the “new hidden morbidity” (Uzark et al., 2008). The new hidden morbidity refers to underlying psychosocial and learning problems experienced by children with cardiac conditions. The identification of psychosocial and learning problems is vital for improving resiliency, quality of life, and comprehensive treatment for children with heart disease (Uzark et al., 2008). Throughout the research on resiliency in children, protective factors have been defined as “specific attributes or situations that are necessary for the process of resilience to occur” (Earvolino-Ramirez, 2007, p. 75). Research on protective factors explores traits that contribute to, influence, facilitate, and change the way an individual responds to trauma (Afifi & MacMillan, 2011). The protective factors identified in most of the research on resilient children are high expectancy, self-determination, positive relationships, social support, self-esteem, self-efficacy, flexibility, and sense of humor (Earvolino-Ramirez). Protective factors such as intelligence, humor, adaptive coping strategies, social connectedness, and social supports are often found within resiliency research (Tusaie & Dyer, 2004).

Resilient children are more likely to have positive relationships with a loving and capable adult, be socially engaging, and be motivated in their academics (Masten, Best, & Garnezy, 1990). An individual’s perception of social and parental support can be both a risk and protective factor; mutual support and understanding between parents and children has been seen as a strong protective factor, but overprotectiveness in parents has

been associated with future maladaptive behaviors and adjustment (Tusaie & Dyer, 2004).

Adolescents' perceived parental support from mother, father, or both parents acts as a protective factor against depression and anxiety; adolescents who endorsed an avoiding coping strategy were more at risk for depression and anxiety (Gomez & McLaren, 2006). Further, an avoidant style of coping when feeling overwhelmed and stressed increased the risk of developing problematic behaviors and mental health concerns (Gomez & McLaren). Risk factors do not predict a poor outcome from adversity or trauma, but rather increase the likelihood of a negative outcome; in contrast, protective factors increase the likelihood of a positive outcome (Tusaie & Dyer, 2004). Additional risk factors noted in the research include poverty, malnutrition, family conflict, and inconsistent parental support, and these risk factors significantly decrease a child's current and future ability to overcome adversity (Masten et al., 1990).

Summary of the literature review.

The diagnosis of LQTS is accompanied by many medical, psychological, and social challenges, resulting in many lifestyle changes and restrictions (Mayo Clinic, 2013). Pediatric health care professionals have identified the challenge of understanding psychosocial and cognitive problems in children diagnosed with LQTS or other congenital cardiac diseases and named it the "new hidden morbidity" (Uzark et al., 2008). Identifying protective and risk factors and strengthening problem solving-abilities are vital to increasing resiliency (Shure, 1994). Resilience is fostered in children in numerous settings such as family, school, peer groups, and religious and athletic organizations in which positive experiences are encouraged and modeled by the behavior

and actions of adults (Masten & Coatsworth, 1998). Masten (2001) describes resilience as not coming from “rare and special qualities, but from the everyday magic of ordinary, normative, human resources in the minds, brains, and bodies of children, in their families and relationships and in their communities” (p. 235).

The goal of this study was to better understand how children with LQTS cope with and manage medical, social, and psychological stressors, in order to further support physicians, psychologists, parents, and others in providing the best treatments, care, and resources needed for improving quality of life and resiliency.

Research question.

What is the experience of children with LQTS with regard to medical care, psychological well-being, and social adjustment?

Chapter 2

Method.

A qualitative research design was used to explore experiences of children with LQTS regarding medical care, psychological well-being, and social adjustment. The following chapter examines use of this research design, inclusion/exclusion criteria, and procedure.

Design.

Kazdin (2003) stated that the main task of qualitative research is to “explicate the ways people in particular settings come to understand, account for, take action, and otherwise manage their day-to-day situations” (p. 333). This study aimed to understand participants’ experiences of living with the life-threatening medical condition of LQTS. Individual interviews provided information about how children at different ages and with different diagnosis histories have lived with LQTS.

Data analysis.

The specific methodology chosen for this study was grounded theory. Grounded theory allows researchers to construct theory from data and information collected from observations, experiences, processes, actions, and interactions from the participants in the study (Creswell, Hanson, Clark Plano, & Morales, 2007). Grounded theory allows the researchers to collect data and information from the participants to generate a hypothesis; this is called hypothesis-generating research (Auerbach & Silverstein, 2003). An important feature of grounded theory is the credibility of the research, meaning how understandable, relevant, and useful is the generated theory to individuals who are in similar situations to those studied (Wagner, Lukassen, & Mahlendorf, 2009).

The grounded theory method of hypothesis generation is known as theoretical coding (Auerbach & Silverstein, 2003). Coding follows a step-by-step process, drawing upon relevant and important texts as well as repeating ideas and themes that are utilized to generate and develop a hypothesis that is considered essential to the research question and phenomenon under study (Auerbach & Silverstein). Open coding, axial coding, and selective coding were the coding strategies utilized in this study. Data analysis started with open coding, where the information is explored in fine detail allowing the data to be compared and contrasted; this generates questions about and insight into the studied phenomena (LaRossa, 2005). Open coding compares and contrasts words, phrases, and sentences called indicators; indicators are then labeled and named as a concept (LaRossa). Next, axial coding is used to make connections from the categories developed during open coding (LaRossa). Axial coding uses the six Cs to further explore and explain the data collected during open coding: cause, consequence, covariance, contingencies, context, and conditions (Aldiabat & Navenec, 2011). Axial coding is focused on explaining the relationships among the variables, categories, and concepts, leading to the generation of hypotheses. Lastly, selective coding is utilized; this groups and connects axial codes into core categories to form the narrative and story of the studied phenomenon (LaRossa). The most salient and relevant variable is called the core variable, which has a relationship with other variables, but is emphasized by the researcher through qualitative, grounded theory research (LaRossa).

Coding was completed by multiple coders, the researcher and two doctoral level graduate students who are members of the LQTS research group at Philadelphia College of Osteopathic Medicine (PCOM). Coders were supervised by a licensed psychologist

with expertise in qualitative research methodology and LQTS. Each coder reviewed the transcripts of the participants, exploring and highlighting relevant text, producing ideas, and generating an initial theory (Auerbach & Silverstein, 2003). In qualitative, grounded methodology, the sample size cannot be determined in advance because the researcher will need to continue interviewing participants and adding new information and concepts to the construction of the theory (Auerbach & Silverstein). The research team will know that they have reached theoretical saturation when new information ends and repeating of information occurs; this signifies that the sample size is sufficient and appropriate (Auerbach & Silverstein). The coding team also needs to have consensual validity, referring to the mutual agreement in assessing the descriptions, themes, and interpretations to produce the most accurate and salient results (Gleelan, 2003).

Participants.

Setting.

Data was collected at the home of seven participants and at Philadelphia College of Osteopathic Medicine (PCOM) for one participant. The interviews were scheduled at a convenient time for the participants and the investigator.

Recruitment.

Children between the ages of 5 and 12 diagnosed with LQTS were recruited to participate in this research study. A letter describing the study was mailed to prospective candidates, along with a return postcard and e-mail address used to confirm interest in participation. Letters and postcards were sent to participants twice, with approximately 6 months between mailings. Information about the study was also sent to local area hospitals and doctors who have contact with prospective participants. Patients who live

in the mid-Atlantic region were also recruited online from informational LQTS websites and other Internet sites, such as Facebook, Twitter, and Craigslist. Additional participants were recruited by word of mouth and snowball sampling. Prospective participants informed the investigator of their interest in the study by either returning the postcard or sending an e-mail. Upon receiving the postcard or e-mail, the investigator called the parent to discuss any questions regarding the study and to arrange a date, time, and location to complete the interviews. All participants who met criteria for this study were recruited from the first mailing of letters and postcards.

Inclusion criteria.

Participants in this study were children in elementary school (Kindergarten to sixth grade), between the ages of 5 and 12, and were diagnosed with LQTS at least 1 year previously. Participants were current patients of an electrocardiologist. Participants were required to have the consent of their parents and/or legal guardians to participate in the study. Participants had to be English speaking. Children of both two-parent and single-parent families were eligible to participate.

Exclusion criteria.

Excluded from this study were children currently hospitalized, those who lost a family member to LQTS within the past year, or those with a serious mental health illness. In this study, serious mental health conditions were defined as any requiring prescription psychotropic medication.

Characteristics.

A personal history questionnaire (Appendix A) was completed by one parent of each participant after signing the informed consent and prior to beginning the interview.

A total of eight interviews were conducted. The children ranged in age from 7 to 12 years old, with the mean age 9.4 years old. This sample consisted of five girls and three boys, seven with LQTS2 and one with LQTS3. Table 1 provides additional family and LQTS information about the participants.

Table 1.

Summary of Demographic and LQTS Information

Pseudonym	Age	Grade in school	LQTS type	Age at symptom onset	Age at diagnosis	Pacemaker or ICD	Mother with LQTS	Father with LQTS	Friends with LQTS
Jillian	9	4	2	N/A	3 months	N/A	No	Yes	No
Matilda	8	3	2	N/A	5.75 years	N/A	No	Yes	No
Lucy	8	3	2	N/A	Birth	N/A	Yes	No	No
Clarrissa	11	6	2	N/A	Birth	N/A	Yes	No	No
Roxy	9	4	3	Birth	Birth	Pacemaker	No	No	No
Benjamin	12	6	2	N/A	7 years	N/A	Yes	No	No
Alexander	7	2	2	6 years	3 years	N/A	No	Yes	No
Richard	11	6	2	N/A	Birth	N/A	Yes	No	No

Note. ICD = implantable cardioverter-defibrillator; N/A = not applicable.

Measures.

A face to face semistructured interview was conducted with each participant. The interview questions (Appendix B) were developmentally appropriate and fostered exploration of the child's knowledge of LQTS and the medical, psychological, and social experiences of living with LQTS.

The Pediatric Quality of Life Inventory (PedsQL) is a self-administered questionnaire designed to measure health-related quality of life in children ages 2 to 18 (Varni, Sherman, Burwinkle, Dickinson, & Dixon, 2004). Participants completed the age-appropriate generic core scales and cardiac modules. Parents of the participants completed the PedsQL parent report forms for the generic core scales, cardiac module, and the family impact module. The child self-report (ages 8-12) and parent report forms of the PedsQL Generic Core Scales are comprised of 23 questions concerning the child's physical, emotional, social, and school functioning (Varni, Seid, Knight, Uzark, & Szer, 2002). The PedsQL Generic Core Scales have demonstrated high validity and reliability (0.88 for the child self-report, 0.90 for the parent proxy-report; Varni, 2013).

The child and parent reports of the PedsQL cardiac modules consist of 27 questions; there are 25 questions on the child and parent reports for young children (ages 5-7). The PedsQL cardiac module explores concerns regarding heart problems, medication, perceived physical appearance, anxiety about medical treatment, and cognitive and communication problems (Gaies, Watnick, Gurney, Bove, & Goldberg, 2001). The PedsQL cardiac modules are utilized with the PedsQL Generic Core Scales and have demonstrated validity and reliability (Gaies et al.; Uzark et al., 2008). The

PedsQL family impact module was completed by the parents and contains 36 questions regarding the impact of the child's health on their own physical, emotional, social, and cognitive functioning, as well as communication problems, feelings of worry, and daily activity and family relationship problems (Varni et al., 2004). This module measures the impact of pediatric chronic health problems on the parent and family, and it has demonstrated reliability and validity (Varni et al., 2004).

Protection of human subjects.

Permission to conduct the study was obtained from the Institutional Review Board of Philadelphia College of Osteopathic Medicine (PCOM).

Procedure.

The parents who were interested in participating in the study were contacted by the investigator via e-mail and by phone. The investigator contacted the prospective participants and discussed the purpose of the study, including the interview and measures. The investigator and parents scheduled a time and location to conduct the interview if the criteria were met. Prior to the interview, the parent reviewed and signed the informed consent form, then completed the personal history questionnaire and PedsQL generic, cardiac, and family impact measures. A semistructured interview asking questions about the child's psychosocial functioning, coping, and management of LQTS was then completed with the children. Interviews with the participants lasted between 16 and 39 minutes. The interview questions were developed, analyzed, and reviewed by the researcher and members of the LQTS research team at PCOM. Each interview session with the child was audiotaped and then transcribed. Transcribing allows information to

be utilized in a usable form for data analysis and ensures that the information gathered in the interviews is correct and consistent.

After the interview, the child completed the PedsQL generic and cardiac modules. The investigator reviewed the transcripts for common themes. A coding scheme was created to define themes that emerged from the information gleaned during the interviews. To reduce error and bias, advanced doctoral students with knowledge and experience in research with patients with LQTS were chosen to review the transcripts and code the data. The investigator and two reviewers met on multiple dates to discuss their impressions of the data and reach consensus on the topics identified. The investigator then interpreted the themes generated from the transcripts and developed a theory relating to these themes. The themes were explored and connected to existing research. Specific quotations from the children in this research study were utilized to best illustrate the meaning of the common themes that emerged and provide a voice for children diagnosed with LQTS. All participants were given two free movie tickets.

Chapter 3

Results.

A qualitative, grounded theory method was utilized to explore how children experience and live with long QT syndrome. Qualitative research is “a process of examining data in order to elicit meaning, gain understanding, and develop empirical knowledge” (Corbin & Strauss, 2008, p. 1). Qualitative research can augment the limited literature and psychosocial research on children diagnosed with LQTS, thereby increasing and enhancing the psychological and social support offered to these children (Marshall & Rossman, 2011).

The major contribution that qualitative research offers is that “our understanding of the experience is elaborated and brought to light in depth as well as in ways that extend our understanding” (Kazdin, 2003, p. 335). A qualitative, grounded theory allows the researcher to value the experiences of the subjects and offers valuable insight and understanding into coping, managing, and living with a serious medical condition (Marshall & Rossman, 2011).

Interviews.

Some of the information about the participants has been changed to protect their identity and to maintain confidentiality. All children were given pseudonyms for this study. The first part of the semistructured interview focused on getting to know the child, and the second part of the interview focused more on their understanding of LQTS. Prior to each interview, parents were asked how they refer to LQTS in their home, if there were any significant changes or events in the past year, and if they had any questions, comments, or concerns before starting the interview. All parents stated that they refer to

LQTS when talking to their child as LQTS. The majority of the parents asked if the interview would discuss sudden cardiac arrest (SCA) or aspects related to dying from LQTS; parents asked the interviewer not to ask any questions related to SCA or death. The interviewer assured parents it would not be broached or asked about during the interview process.

Initial impressions.

After a short discussion with the parents, the researcher spoke to each child about the basics of the interview and offered praise and appreciation for being a participant in the study. Some of the children discussed their anxiety, while others exhibited anxiety by leg shaking, nail biting, and avoidant behaviors and answers. The interviews started with the “getting to know you” section about family, friends, school, classes, likes, and dislikes. The second part of the interview focused on LQTS related questions. As the interview process continued, the children seemed to feel more comfortable with the interviewer and questions. The researcher believed that the children seemed more nervous when asked questions about LQTS, and the researcher offered encouragement and praise during times that seemed difficult for the children.

Living with LQTS: A child’s perspective

The children in this study all had at least one sibling, with the exception of one participant. Three had siblings diagnosed with LQTS, and all but one participant had a parent diagnosed with LQTS. In addition to the themes that will be described below pertaining to LQTS, the children had normal, age-appropriate concerns and described managing school, frustrations with homework, excitement about sleepovers, having fun with friends, and loving their pets. The children in the study also discussed plans for

their future, such as getting a perfect score on the SAT and career goals. The children described and explored their relationships with their parents, siblings, friends, and peers. Overall, these children appeared comparable to children not diagnosed with LQTS with regard to their interests, activities, and concerns.

Study population.

Table 2 shows the sample size, mean, range, and standard deviation data for the children who completed the Child Report of the PedsQL 4.0 Generic Core Scale and PedsQL 3.0 Cardiac Module. The PedsQL measures quality of life by asking children and parents how much of a problem they are having in areas of physical, social, emotional, school, and family functioning. Responses to the questions are *never* (100), *almost never* (75), *sometimes* (50), *often* (25), and *almost always* (0), with higher scores indicating a higher quality of life for children (Varni, Limbers, & Burwinkle, 2007). Average scores ranged from 73.75 to 88.75, with the lowest averages found in perception of school functioning and treatment anxiety. Higher scores were attributed to the children's perception of their social functioning and perceived physical appearance. The children reported high quality of life on the generic and cardiac domains, as evidenced by average scores above 80 on more than half of the subscales. However, these scores were lower than those of healthy children, whose scores frequently range between 83 and 100 (Varni, Seid, & Kurtin, 2001).

Table 2.

Child Self-Report PedsQL 4.0 Generic Core Scale and PedsQL 3.0 Cardiac Module Scores

Scale or Dimension	<i>X</i>	Range	<i>SD</i>
Physical functioning	85.21	71.88-93.75	7.73
Emotional functioning	80.00	50.00-95.00	17.32
Social functioning	88.75	80.00-100.00	8.76
School functioning	73.75	45.00-80.00	13.82
Psychosocial health summary	80.84	66.67-91.67	8.91
Physical health summary	85.21	71.88-93.75	7.73
Total Generic Core Scale score	78.99	67.39-92.39	12.11
Heart problems and treatment	83.03	71.43-96.42	7.81
Treatment II	87.71	66.67-95.00	10.19
Perceived physical appearance	88.54	66.67-100.00	11.73
Treatment anxiety	75.00	37.50-100.00	18.60
Cognitive problems	78.13	70.00-90.00	7.99
Communication	80.21	58.33-100.00	18.33
Total cardiac module score	82.11	68.00-93.52	9.05

Table 3 shows the sample size, mean, range, and standard deviation data for parents of children with LQTS who completed the Parent-Proxy PedsQL 4.0 Generic Core Scale and PedsQL 3.0 cardiac module. Average scores ranged from 71.67 to 95.67. The lowest scores were attributed to the perception of their child's communication and treatment anxiety. Higher scores were found for perception of treatment barriers and the child's perceived physical appearance. Parents reported high quality of life on the generic and cardiac domains, as evidenced by average scores above 80 on more than half of the subscales (Varni et al., 2001).

Table 3.

Parent-Proxy Report PedsQL 4.0 Generic Core Scale and PedsQL 3.0 Cardiac Module Scores

Scale or Dimension	<i>X</i>	Range	<i>SD</i>
Physical functioning	85.00	71.88-100.00	10.08
Emotional functioning	75.00	50.00-90.00	14.34
Social functioning	88.50	75.00-100.00	11.56
School functioning	80.50	50.00-100.00	19.50
Psychosocial health summary	81.33	53.33-91.67	10.68
Physical health summary	85.00	71.88-100.00	10.08
Total Generic Core Scale score	82.60	60.87-90.12	8.89
Heart problems and treatment	87.50	82.14-92.86	3.86
Treatment II	95.67	90.00-100.00	4.17
Perceived physical appearance	89.17	66.67-100.00	15.74
Treatment anxiety	73.75	50.00-100.00	19.72
Cognitive problems	78.50	75.00-100.00	14.35
Communication	71.67	33.33-100.00	23.31
Total cardiac module score	83.61	73.15-92.59	7.66

Table 4 shows the sample size, mean, range, and standard deviation data for parents who completed the PedsQL 4.0 Family Impact Module. Average scores ranged from 68.13 to 87.21. Lower scores were attributed to the parents' perception of worry and communication. Higher scores were found for the parents' perception of physical functioning and cognitive functioning. The data revealed that parents reported difficulties in the domains measuring emotional, social, communication, worry, family daily activities, and relationships, as evidenced by average scores below 80 on more than half of the subscales (Varni et al., 2001).

Table 4.

PedsQL 4.0 Family Impact Module

Dimension	<i>X</i>	Range	<i>SD</i>
Physical functioning	87.21	65.20-100.00	14.55
Emotional functioning	78.75	45.00-100.00	20.66
Social functioning	78.13	31.25-100.00	25.00
Cognitive functioning	83.13	45.00-100.00	23.89
Communication	70.83	25.00-91.67	23.15
Worry	68.13	45.00-85.00	14.62
Daily activities	76.04	25.00-100.00	33.76
Family relationships	79.38	50.00-100.00	21.45
Parent HRQL	81.88	47.50-100.00	18.55
Family functioning	78.13	50.00-100.00	19.41
Total score	78.21	50.00-95.14	16.00

The PedsQL scores also demonstrated that 6 of 8 children and 6 of 10 parents had scores below 83 on the domains of cognitive problems and treatment anxiety, whereas scores for healthy children usually range between 83 and 100 (Varni et al., 2001). The

cognitive problems domain assessed difficulties with problem solving, attention, and memory; the treatment anxiety domain assessed fears related to medical treatments, having to go to the doctor or hospital, and waiting to see the doctor. There were differences in that parents reported difficulties in the domains of emotional functioning and communication, with scores below 83, whereas children had higher quality of life scores within these domains. Also, all the children reported more difficulties in the school functioning domain, with scores below 83, whereas 5 of 10 parents had scores above 83; scores in this domain for healthy children typically range from 83 to 100.

Chapter 4

Discussion

Overall, the PedsQL findings of this study indicate that the children and parents have a positive perception of the child's ability to manage and cope with LQTS. However, these findings suggest more difficulty managing and coping as a family, as indicated by the PedsQL family impact module scores. Lower scores on the family impact module than on the PedsQL child and parent reports for both generic and cardiac modules may indicate that parents are sacrificing self-care for the care of their children, as well as the parents' anxiety, worry, and fear of the unknown regarding LQTS diagnosis and treatment.

Developmental stage.

The children in this study were between the ages of 7 and 12 years and, according to Erickson's developmental model, are in the stage of industry versus inferiority. Erickson believed this stage was crucial for developing and strengthening social skills and self-esteem (Sigelman & Rider, 2009). Peers and teachers become more important to children as they strive to develop new skills, take pride in their accomplishments, and work toward the virtue of competence (Sigelman & Rider). Children in this study shared their thoughts, feelings, and insights about being a preteen diagnosed with long QT syndrome, along with the stress and anxiety that accompany being a preteen. The children discussed various ways of coping and managing stress and anxiety, including the use of what could be described as cognitive behavioral strategies.

Cognitive behavioral strategy of coping.

Individuals who are feeling depressed and/or anxious may engage in the coping strategy of avoidance (Beck, 2011). Avoidance was observed in statements that denied the physical and lifestyle limitations for a child diagnosed with LQTS, such as “I can do everything” and “it doesn’t limit you at all.” Another avoidant coping strategy used by some of the children was saying “I don’t care,” which may have masked feelings of worry and sadness about the diagnosis and treatment of LQTS. Humor and silly behavior were also seen during the interviews and may have been a means to avoid uncomfortable thoughts and discussion about LQTS.

Magical power.

Four of the eight children stated that if they were able to have a magical power, they wished they could fly. Two of the children described why they would like to fly, stating “I could see, like, stuff on the ground that, like, I couldn’t see before” and “flying is a cool thing . . . because, like, you can get there without a car, gas, or anything.” Another child described being able to fly as feeling more free. Three of the eight children discussed being able to change their shape with one child discussing the magical power of shape shifting “because you could turn into anything you wanted to be, and you could even, like, turn invisible because you could shape shift into, like, anything so you could basically do anything you wanted.” One child discussed the magical power of being invisible or pausing time, and another child discussed being able to be incredibly strong and to be amazing at sports.

Themes.

There were several themes revealed in the data that were expressed by the children during the interviews. In this study the main themes were the treatment of LQTS, their relationships, and social connectedness.

Treatment.

Several topics about treating LQTS surfaced during the interviews. Treatment included discussion of the stress test, the use of medication, hospital setting, lifestyle restrictions, and the doctor-patient relationship.

Stress test.

The stress test was discussed by most of the participants in the study. When discussing visiting the doctors, the participants described the experience of engaging in the stress test. The children discussed both positive and negative experiences of engaging in the stress test which, depending on age, utilizes a treadmill or bike. One child stated “they put me on a bike or treadmill and see how high my heart rate goes on that.” Another child described goals of getting to a higher levels on the treadmill; “My goal is to try to beat the 8-year-olds.” One child discussed using the bike instead of the treadmill, while another child stated that she does not like taking the stress test on her doctor visits. Seemingly, the children in this study have found a way to cope with this particular test.

Medication.

All of the children in the study were prescribed medication for treating LQTS and expressed that the medicine was a vital part of treatment. They also discussed that they take medication to treat their LQTS multiple times a day. All the participants discussed needing to take their LQTS medication to keep their heart healthy. Medication

compliance is crucial in treating LQTS, lowering the risk of a cardiac event when the medication is taken on a consistent basis (SADS Foundation, 2008). Most of the children discussed taking the medication at home; however, some of the children discussed taking medication at the school if it's forgotten in the morning, and one participant is scheduled to take it at school. One child described being questioned by a peer about her medication and responding "it's just for my heart." Another child stated that she was happy that her medication was recently changed from a liquid to a pill "so that it is easier for me to take." One participant described "feeling weird" having to take his medication if friends are visiting.

Going to the hospital.

The children described feeling awkward, nervous, and weird, disliking or being impartial to going to doctor appointments. They described feeling "a little awkward because, I mean, I'm the only kid in my school that has this," and "a little weird that I am going to the hospital for a doctor's appointment." One child also expressed her fear and anxiety regarding something potentially going wrong during the doctor's appointment. Other children seemed to have neutral feelings about going to the doctor, whereas some did not enjoy going, but understood the benefits of their medical appointments for LQTS. One child engaged in distracting behaviors when asked about going to the doctor appointments.

Doctor-patient relationship.

The children in this study described the importance of a positive relationship with their doctor. The doctor being "nice" was described as a positive characteristic of good treatment for the children. Another positive aspect in the doctor-patient relationship is

the competence of the doctor. One child discussed that her doctor “is, like, the best heart doctor in the world . . . so that makes me feel a little safer.” She further stated “so that I know she is, like, a good doctor so she will kind of keep me going, I guess.”

Lifestyle restrictions.

As part of treatment, children diagnosed with LQTS have lifestyle modifications that restrict or limit their diet and engagement in physical activities. These restrictions are necessary to avoid triggering a cardiac event (Farnsworth et al., 2006). However, not being able to participate with their peers in the restricted activities may have a negative impact on relationships with peers, self-esteem, and acceptance of LQTS (Vetter, 2007). The children in this study discussed activities, sports, foods, and beverages that they are limited or not able to partake in due to their treatment of LQTS.

Limitation of physical activities in the home, community, and school settings were discussed by most of the children. Soccer, track, swimming, basketball, and field hockey were sports that the children stated they were not able to play. The children also discussed limitations during gym class in school, with one child stating “I really like gym, but I can’t do that much because of my heart condition.” Another child discussed not being able to participate in physical activities (pushups, sit-ups, rock climbing) or how she sometimes needs assistance from the gym teacher. One child explained a school fitness test called the mile run where the child had to walk the mile, “Everybody else did it in 6 minutes because they could run, and I had to walk and I did it in 13 minutes.”

Some of the children discussed abstaining from physical activities, but did not state that it was because they were not allowed to or restricted. Two of the children discussed that they used to play soccer, but they do not play anymore. When asked why

they no longer play, one child stated “I don’t know, maybe I just didn’t like it, I don’t remember” and the other child stated “Um, my mom stopped signing me up for it.” Two of the children strongly expressed that they are not physically limited or restricted by LQTS. One of the children stated several times during the interview that LQTS “doesn’t really limit you,” and the other child stated “I can do everything.”

In addition to physical activity restrictions, the children in this study discussed food and beverage restrictions. They discussed not being able to have caffeine or soda and having limits with candy. One child described her wish to try chocolate, “Um, one time, what I would want to do is, like, I would just want to try chocolate . . . but I know I can’t have that, but I want to know how it will taste to see if I would like it.”

Relationships.

Parent-Child relationship.

Several aspects of the parent and child relationship surfaced throughout the study. All of the children discussed that their parents are active and involved with their medication management, attending doctor’s appointments, and overseeing the treatment regimens.

Research has shown that parents of children diagnosed with LQTS are active in seeking information regarding treatment, medication, surgery, and ways of educating others about LQTS (Burns-Pentecost, 2013). However, how much information should be given to the child is often a very difficult decision; parents walk a fine line between offering information regarding LQTS and creating a sense of worry and fear. One child discussed her belief that she has a limited understanding of what LQTS is by saying “I don’t really know that much because I’m still little, and my mom hasn’t told me that

much because she doesn't want me to worry." Other children described LQTS as being heart related. Two children were able to express more detailed information about LQTS. One child discussed that "not many people die from it, but you can die if you don't figure it out," "it's not contagious or anything, you have to be born with it" and that it can be "pretty deadly if you don't have your pacemaker." The other child explained that "it's a defect in your heart" and "our hearts are slower than most."

The children in this study discussed positive and negative aspects of the concerns and worry expressed by their parents. Some children viewed their parents' worry in a positive manner. Some of the children expressed that their parents' worry made them feel safe and expressed positive feelings about the attention they were receiving. Others felt that their parents did not worry about them. One child discussed that her grandfather is a doctor and explained "I am kind of, like, in good hands and I am OK now . . . he doesn't worry, so then everyone else knows I am ok." Another child discussed the difference in the worry between her parents, explaining that her father worries because he does not have the diagnosis of LQTS, whereas her mother who has the diagnosis worries also, but "it's not that big of a deal." Some described their parents' worry in an undesirable light. They discussed frustrations with reminders regarding the LQTS treatment, sadness due to the parents worrying, and avoiding the topic of the parents' worry.

Animals.

Another theme with all eight children was that they all lived with an animal (dog or cat). Also, three of the children take horseback riding lessons and love grooming

horses. Some of the children also discussed playing with their dogs, with two children wishing for more dogs in their family.

Friendships.

The importance of friendship was discussed by all eight participants during the interviews. Every child in this study had a group of friends, and each child discussed having at least one best friend. The children discussed engaging in activities and events with friends such as sleepovers, sports, and other typical social activities. Children in this study varied in how they viewed sharing information about LQTS with friends. Some discussed ambivalence about talking to friends, while others try to keep the information private.

Some of the children discussed gratitude and appreciation for being able to share with their close friends their thoughts and feeling about LQTS. These children discussed sharing personal information with friends and finding comfort that their friends seem concerned and interested about the diagnosis and treatment of LQTS.

Other children in this study preferred to keep their diagnosis private. The children with this preference all acknowledged that their friends know they have a heart condition, but do not discuss LQTS with them. These children discussed keeping their LQTS information more confidential because of fears of more peers and parents finding out about the LQTS diagnosis. Other children expressed not wanting to share that they have LQTS and keeping it more personal, stating “‘cause I don’t really think it’s any of their business to, like, know about my thing.” Another child stated “like, they know about it, but I don’t talk to them about it.” He stated his friends will ask questions “like what can

and can't you do." When asked about discussing LQTS with friends, one child emphatically stated no, and the other responded "not really."

The children in the study discussed numerous values and qualities that they like about their friends. Overall, the children expressed that their friends have similar interests to theirs, are fun, friendly, and nice, and have a good personality. The children also described important qualities in their close friends as being trustworthy, strong, and having the ability to "cheer me up" as important.

Social connectedness.

Stigma.

The children in this study also discussed some fears of being viewed differently or not accepted by their peers.

Diagnosis.

The children in this study discussed positive feelings about their teachers knowing they have the diagnosis of LQTS to help keep them safe, but discussed some ambivalence and worry related to their peers knowing about their diagnosis. Some of the children discussed that they were not bothered by teachers and peers knowing their diagnosis, with two of the children stating, "I don't really care because, I mean, for my safety they need to know," and the other stating that she doesn't "mind everybody knowing about the LQTS diagnosis." Other children discussed fear of being teased by their peers and feeling uncomfortable.

Holter monitor.

The use of Holter monitors that measure and record heart rate was discussed by the children in this study. Some of the children expressed feelings of embarrassment and

being physically uncomfortable wearing the Holter monitors. One child discussed hiding the heart monitor in school with a sweatshirt, and another explained that he felt that the monitor negatively affected him while playing baseball. The children also discussed explanations that they give to peers who ask about the heart monitor. Some of the children responded to peers with short answers like “the doctor says I have to wear it” and “it’s none of your business.” Other children discussed explaining the heart monitor to their peers. “I kind of tell them it’s, like, a phone for my doctor, to make sure that she knows that everything is OK with me” and “this is my pager...it monitors my heart.”

Feeling sad and lonely.

The children in this study expressed feelings of sadness, being different, and lonely. They expressed not knowing other children who are diagnosed with LQTS and reasons why they would like to meet another child their age with the LQTS diagnosis. Some of the children stated that they feel sad and wish they didn’t have the diagnosis of LQTS. One child stated that it makes him feel “kinda stressed...although the stress, I’m not sure, maybe it’s just sadness.” The children also discussed feeling different than their peers and wanting to meet another child with LQTS with one child stating “to feel like I am not the only kid who has it.”

Theoretical analysis of findings.

In this study, how children experience and live with long QT syndrome was examined. Previous research has explored how parents cope with their children’s diagnosis of LQTS and manage their quality of life, anxiety, fears, worry and potential issues of loss resulting from LQTS (Burns-Pentecost, 2013; Gonzales, 2009; Haynes-Weller, 2011; Janney, 2011). In this study, the children discussed and expressed their

experiences and identified factors that can be seen in the “Five Pillars of Adaptation for Long QT Syndrome” that influence how they cope with and manage living with LQTS.

The five pillars of adaptation for long QT syndrome.

A *pillar* is defined as being a firm upright support, an integral and upstanding member or part, and a fundamental precept (Merriam-Webster, 2015). The Five Pillars of Adaptation for Long QT Syndrome were born from this research. They are the fundamental elements through which we can learn how a child lives with long QT syndrome. The Five Pillars of Adaptation for Long QT syndrome are described below.

Pillar 1: Developmental level and self-esteem

The children in this study described experiences such as playing with friends, frustrations with school regarding homework, their friends, family, love for their pets, hobbies, and other normal, age-appropriate activities and interactions. According to Erickson, children move through stages, learning and developing skills and competencies (Sigelman & Rider, 2012). During the industry versus inferiority stage, children ages 6 to 12 are usually in school, and peers and teachers become more crucial in developing and strengthening social and cognitive skills (Sigelman & Rider). In this stage, children start to experience physical, social, and emotional changes that may impact their cognitive and emotional level. These times for children can be very challenging, even more so for a child with LQTS because their diagnosis may be an additional social stressor. The children in this study indicated on the PedsQL difficulties in the cognitive, treatment anxiety, and school domains. Their developmental level may be a significant factor in these difficulties as they become more aware of the implications of the diagnosis and treatment of LQTS and find a balance between managing their social relationships

and school demands (Gallagher, 2014). Children diagnosed with LQTS may encounter questions about their condition from their peers and have lifestyle restrictions that prevent them from fully participating in school or social activities.

During the preteen stage, children start to evaluate and compare themselves to their peers (McAdams & Olson, 2010). Individual differences emerge, and for most children, self-esteem begins to decrease compared to self-esteem prior to the age of 7 (McAdams & Olson). As children continue their development, parents of children diagnosed with LQTS may struggle with how much LQTS information to give their children without overwhelming them. Gonzales (2009) found that parents struggle with communicating with their children about LQTS, depending on the child's personal and social development. Seven of eight parents in the present study asked that the interviewer not discuss or explore sudden cardiac arrest (SCA) with their children. Parents want to protect their children from fear, anxiety, and worry related to the LQTS diagnosis (Burns-Pentecost, 2013).

Strategies of avoidant coping are used to protect an individual from anxiety (Beck, 2011). Verbal avoidant coping strategies, such as "I don't care," "it doesn't limit me," or "I can do everything," were expressed when exploring the lifestyle impact of LQTS. These statements can be a way for children to distance themselves from the reality of LQTS and the attendant physical limitations and lifestyle restrictions. The statements may also mask genuine feelings the children have, such as sadness, anxiety, or loneliness (Whitson, 2013). Children may use other avoidant coping strategies, such as engaging in silly or distracting behaviors, to avoid negative thoughts and feelings related to LQTS.

Pillar 2: Peer and social relationships.

Pillar 2 explores peer relationships, the role that friends play for a child diagnosed with LQTS, and fears and worry about bullying and being ostracized for being different. Children diagnosed with LQTS may face more challenges than healthy peers, due to the physical restrictions on sports, diet, social opportunities, and the feelings of loneliness and being different (Martinez, Carter, & Legato, 2011). The children in the study all discussed having close friends and engaging in normal, fun-filled activities with their friends. Developmentally, friendships and relationships with peers are extremely important. Research has shown that children with a chronic disease or disorder, such as LQTS, achieve better adjustment when they have high quality friendships and acceptance by peers (Martinez, Carter, & Legato).

Children sharing their diagnosis with others are at higher risk for social alienation and bullying (Watson, 2011). Children in this study identified feelings of being different and fears of being treated negatively or bullied by the “mean kids.” One child stated “I’m not really happy the students know ‘cause then they’ll be, like, hey look. I know what’s happening to her . . . I think only teachers should know, none of the kids” and also “I only tell the nice kids. The mean kids I don’t tell.” Another child stated “sometimes I feel a little bit different than other kids” describing feelings about having LQTS. The children in the study were able to balance the fear of being picked on or viewed differently by their peers with the acceptance and support by their close friends. The emotional support children with LQTS receive from friends can serve as a protective factor against social isolation (Martinez, Carter, & Legato, 2011).

Pillar 3: Parental support.

Pillar 3 examines how the parent-child relationship influences the child's ability to manage and cope with the diagnosis and treatment of LQTS. The children in the current study all discussed the lifestyle changes, restrictions, and worry their parents have for them. The children shared their thoughts on how their parents monitor their physical activities, diet, medication, and treatment. Parental perceptions factor significantly into how children cope with LQTS. Researchers have found that pressured vigilance by parents and others was frustrating, and not being treated differently from other peers was also important to children (Kendall, Sloper, Lewin, & Parsons, 2003). In a study exploring experiences of parents who have a child or children diagnosed with LQTS, parents described difficult lifestyle-restricting decisions they have made for their children to keep them safe and protect them from triggers, as well as a pressure to create for their children a fun and stress-free life (Farnsworth et al., 2006). Most parents expressed anxiety and fear related to LQTS and described many lifestyle changes made to alleviate their fears and reduce the chance of sudden death due to LQTS (Farnsworth et al.). Parents described fear-alleviating strategies, such as consistent vigilance with their children and providing education about LQTS to those in medical, social, and academic settings (Farnsworth et al.).

In a study of parents with children who are at risk for sudden cardiac death, perceptions their child's quality of life are influenced by stress related to caring for their at-risk child and their own mental health state, welfare, and responsibilities (Smets et al., 2008). However, parents were able to give an appropriate assessment of their child's psychological and emotional well-being, without allowing their own concerns and

anxiety to dominate and without overestimating their child's capabilities (Smets et al.). Further, their ability to do so comforts the child and decreases the child's worry (Smets et al.). Although the children in this study discussed frustrations with vigilance from parents and with restrictions, they also expressed positive thoughts and feelings regarding their parents worry about them, stating that they feel "safer" and "feeling pretty good because my family is aware of me." One participant stated his frustration with parental hypervigilance, but added "I know it's the right thing."

Parents of children who have inherited cardiovascular diseases reported feeling unsure of the social and emotional assessment of their children compared to their perceptions of physical assessment (Smets et al., 2008). In another study, parents with children who have congenital cardiac disease saw a need for more information about heart disease and the appropriate distribution of the information to communities and schools; however, parents struggled with anxiety that the information may cause their children to be treated differently or as special due to their medical condition (Kendall et al., 2003). The parents agreed that having the child lead a normal life was important, but they needed to understand which activities and situations put the child at most risk for a cardiac event (Kendall et al.). Most parents in this study were concerned with their children's knowledge that LQTS can have fatal consequences, wanting to shield them from information that may be emotionally overwhelming. Parents may struggle with how much information about LQTS is too much or not enough for their children. In the current study, most of the children knew the basics of LQTS; they knew that it is a heart condition and treatment consists of lifestyle modifications and medication. One participant discussed limited knowledge due to her age and her parent not wanting to

worry her. Zolten and Long (2006) explored various ways to support parents with children who have a chronic illness, such as explaining the diagnosis to the child in an open and honest way, that is not their fault, and encouraging the child to express his or her thoughts and feelings about LQTS, diagnosis, and treatment.

Pillar 4: Social problem solving.

Pillar 4 explores how the child with the diagnosis of LQTS engages in social problem-solving in school and social situations. The social problem solving model (SPSM) is a multidimensional model that helps individuals recognize, develop, and strengthen adaptive coping strategies for the problems in their lives (Nezu & Nezu, 2013). The social problem solving model helps individuals become more goal focused, overcome barriers and conflicts from achieving their goals, and increase adaptive emotional reactions to the problem (Nezu & Nezu, 2013). The model also explores the individual's perception of the problem, ability to cope with the problem, the problem-solving style utilized when attempting to alleviate the problem, stress, and ability to find a solution (Nezu & Nezu, 2013). The social problem-solving model can help children gain insight and understanding into how to cope with and adjust to the stress and anxiety that may accompany their LQTS diagnosis and treatment.

Within a problem-solving approach, effective problem solvers have an increased chance of exhibiting resiliency after experiencing negative life events and significant stressors, whereas ineffective problem solvers are more likely to experience psychological concern (Nezu, Nezu, Felgoise, McClure, & Houts, 2003). For example, a child who is living with LQTS and is an effective problem solver may exhibit a higher level of adjustment and less distress than a child who is less effective at problem solving.

In addition, having strong problem-solving skills has been found to be effective in increasing the quality of life among parents and caregivers of individuals with chronic health conditions (Nezu, Nezu, & D’Zurilla, 2013). A child’s problem-solving ability is crucial to helping solve and resolve problems in life, as well as to improving ability to cope with, manage, and overcome stressful situations (Shure, 1994). Effective problem solving for the children in this study was evidenced by being able to explain to their peers the LQTS diagnosis and reasons for the heart monitor and manage distressing feelings about having LQTS. In addition, some of the children were able to explain to their friends and peers some of their lifestyle changes and physical restrictions due to their LQTS. The use of effective problem solving has been seen to decrease stress, anxiety, and worry (Burns-Pentecost, 2013; Janney, 2011).

Children diagnosed with LQTS who are having difficulty coping with the diagnosis and treatment of LQTS can benefit from engaging in problem-solving therapy (PST). Problem-solving therapy is a cognitive-behavioral intervention designed to enhance quality of life and coping skills through development and strengthening of problem-solving abilities, attitudes, and skills (Nezu, Nezu, & D’Zurilla, 2010). The major goal of PST is to help individuals identify and solve problems in their lives while teaching them problem-solving skills to use for future problems and stressors (Nezu, Nezu, & D’Zurilla, 2013). In addition, children who may be struggling to adjust to the diagnosis and treatment of LQTS may benefit from participating in programs designed to foster healthier relationships with peers, prosocial behaviors, and problem-solving skills (Shure, 2000).

Shure (2000) developed a program called I Can Problem Solve (ICPS) for children ages 4 to 7 and preteens ages 8 to 12. The ICPS program for children ages 4 to 7 focuses on developing and increasing the child's problem-solving ability and conflict resolution skills, in addition to identifying and understanding their own and others feelings (Shure, 1994). Problem solving can support the children in identifying their true and genuine thoughts and feelings related to LQTS as they work towards lessening avoidant coping strategies used to reduce their stress and anxiety. The ICPS program helps children increase their ability to think for themselves and solve problems, as well as increasing their self-esteem, confidence, and competence (Shure, 2000). Parents can also learn skills to foster problem-solving ability and increase communication with their children, providing the groundwork for a well-adjusted, motivated, and competent teenager (Shure, 2000). Children diagnosed with LQTS who can effectively problem solve are more likely to exhibit resiliency and the ability to successfully cope with and manage physical, social, or emotional stressors related to their diagnosis and treatment (Shure & Aberson, 2006).

Pillar 5: Treatment and resources.

Pillar 5 explores the treatment of LQTS and social resources that may increase the child's social connectedness. From a medical standpoint, the treatment of LQTS can consist of medications, lifestyle changes, surgery, and medical devices (Mayo Clinic, 2012). Parents in this study wanted the best possible treatment and care for their child. However, the children in this study discussed different aspects of their treatment that they thought were important for them. As parents value clear, concise, and direct communication with them by the physician regarding the LQTS diagnosis and treatment

(Steinhauser, 2010), the children in this study valued a physician who is “nice” to them and a place where they can feel safe. The children also discussed “feeling different” because of going to a hospital for their doctor appointments and frustrations with medication, stress test, and heart monitor. The children in this study either voiced their frustrations about the lifestyle modifications and restrictions due to the LQTS diagnosis or tried to deny that LQTS is limiting.

One way to counter this new hidden morbidity is to bring these psychosocial concerns into the open. In this study, when asked about what they would say to a friend who had to go to the heart doctor, the children responded with reassuring statements, such as “don’t be nervous,” “everything will be all right,” and “don’t be afraid.” One child stated “I would say the next day after she had it, how do you feel about that, did it scare you, how did it make you feel?” Reassurance, encouragement, and exploring thoughts and feelings about the doctor’s visit and treatment were valued by the children in this study. As explored in pillar 4, children, parents, and families affected by LQTS are encouraged to engage in and increase problem solving skills. The social problem-solving model has been shown to be effective in increasing abilities to recognize problems and solve them and in decreasing symptoms of depression and anxiety (Eccleston, Palermo, Fisher, & Law, 2013).

Children who are diagnosed with LQTS are able to make changes, such as restrictions in exercise and sports, taking medication, and in some cases, having medical devices and surgical procedures, to prevent an onset of symptoms (Mayo Clinic, 2013). Children in this study described difficulty managing the restrictions on their physical activity and diet. Individuals who are diagnosed during adolescence may have more

trouble adjusting to the LQTS lifestyle changes than children diagnosed at a younger age because those diagnosed younger are more accustomed to living with LQTS (Farnsworth et al., 2006). Some of the children in this study described initial feelings of being nervous and anxious about LQTS, but stated that they are used to it, it doesn't bother them anymore, or they do not care.

One message echoed by each child was the thought of being different. Children stated they were “the only one” who has the LQTS diagnosis and wanted to meet another child their age who has the diagnosis of LQTS. Currently, LQTS support groups for children are lacking. Social media LQTS support groups are available for parents of children with LQTS. Burns-Pentecost (2013) conducted a study with parents coping with their children's LQTS diagnosis using an online user group. Similar to the parents in the present study, parents in the research by Burns-Pentecost described feelings of being overwhelmed, worried, frustrated, stressed, and isolated (Burns-Pentecost). Peer social support using the social media was effective for the parents with seeking information, advice, support, and connections with group members (Burns-Pentecost). Parents expressed being thankful and being grateful for the LQTS group (Burns-Pentecost). Support groups for individuals diagnosed with a chronic illness can help lessen feelings of loneliness, depression, and anxiety (Mayo Clinic, 2015). These groups can also help foster and increase a sense of control, empowerment, sense of adjustment, and coping skills (Mayo Clinic, 2015). With facilitation by a trained therapist with knowledge of LQTS, children diagnosed with LQTS can receive these benefits and have opportunities to express their genuine feelings, attain more social resources, and gain a clear understanding of and insight into current and future situations (Mayo Clinic, 2015).

The five pillars of adaptation describe the importance of understanding the child's developmental level, relationships with peers and parents, social problem-solving ability, and aspects of treatments and resources needed to successfully manage and cope with the diagnosis of LQTS. These pillars provide a means and framework for supporting a child with managing the diagnosis and treatment of LQTS. The five pillars are also designed to help the child balance normal developmental concerns, along with the challenges that accompany having LQTS. The pillars can also help increase self-esteem, self-efficacy, problem-solving ability, and resilience, decreasing stress, anxiety, and fear of the unknown.

Happiness, as compared to stress, has been found to lower the risk of having a cardiac event (Lane, Reis, Peterson, Zareba, & Moss, 2009). Positive emotions such as happiness are a protective factor associated with reduced risk of susceptibility to a cardiac event and may indicate that the negative emotions suffered by a child diagnosed with LQTS increase the risk of a sudden cardiac event (Lane et al). Further, positive emotions and engagement in behaviors and activities that enhance the likelihood of experiencing a positive emotion should be utilized within the planned treatment approach for an individual with a serious medical concern such as long QT syndrome (Lane et al.). Support, love, and understanding were important for children adjusting to and living with congenital cardiac heart conditions (Kendall et al., 2003), as also found with the children in this study.

Limitations.

The sample size for this study was small and homogenous. In addition, three of the eight participants were siblings diagnosed with LQTS. With these limitations,

generalizability of the findings is very limited, due to lack of diverse representation and the small number of participants. Another limitation of the study is that the participants and their parents volunteered for the study and may represent children diagnosed with LQTS and their parents who already have developed the qualities needed to manage and cope with the LQTS diagnosis in a healthy and supportive way. All eight children chose to complete the interviews and were supported and encouraged by their parents. The children in the study had a choice to decline participation, and all eight chose to engage in the study. The children and their parents demonstrated acts of altruism to help other children and families diagnosed with LQTS.

The researcher conducted face to face semistructured interviews with the participants, primarily in their homes, and started the interviews with praise and encouragement. Thus, there may have been self-report or social desirability bias, in which children may have answered questions in the way that they thought the researcher wanted them to answer, or unintended bias, as the researcher offered the children praise prior to and during each interview (Auerbach & Silverstein, 2003).

Similar to quantitative research, problems with internal validity, consensual validity, and interrater reliability can occur with qualitative research (Barbour, 2001). This study used multiple coders to increase interrater reliability and improve frameworks and interpretations, while decreasing subjectivity in the data analysis (Barbour). Despite using multiple coders to increase interrater reliability and ensure consensual validity among emerging themes and patterns from the transcribed text, analysis of the data is subject to human error and experimenter bias.

Replication of study.

If the researcher were able to conduct this study again, several changes would be made. First, there were many potential participants who wanted to be part of the study, but lived outside the geographic area that was one of the inclusion criteria. Being able to include these subjects by increasing the geographic boundaries would increase the sample size, improve generalizability, and contribute additional information. One method of achieving this would be to utilize a webcam or cell phone camera to conduct interviews with children outside the local area. Also, during transcription, the researcher identified lost opportunities to engage in more in-depth conversations on certain topics. More insight and understanding may have been gained by having a follow-up interview with the participants.

Future directions.

The current study provided insight into the experiences and quality of life of children living with LQTS. The information gathered here can provide insight into and understanding of the protective factors, risk factors, and needs for support for a child with LQTS throughout his or her development. In addition, the five pillars of adaptation for LQTS highlighted the important influence of understanding the child's developmental level, relationships, social problem-solving ability, and LQTS treatment and the social resources needed to successfully manage and cope with LQTS.

These findings may also add to the research on the psychosocial effects of living with LQTS and how children manage and cope with this and other serious medical conditions. This study has expanded upon the existing literature pertaining to LQTS by providing more understanding of the effects of the disease and how a preteen child

interprets, perceives, manages, and copes living with LQTS on a daily basis. Future researchers also may benefit by comparing the differences and similarities of young children, preteens, and adolescents managing and coping with the diagnosis of LQTS.

These findings are projected to lead to more public awareness of LQTS and improvements in treatment, support, and psychoeducation for children with LQTS.

Specifically, support groups for children diagnosed with LQTS are currently lacking and needed, as voiced by the children in this study. Future research on the benefits of LQTS support groups is encouraged.

References

- Afifi, T. O., & MacMillan, H. L. (2011). Resilience following child maltreatment: A review of protective factors. *Canadian Journal of Psychiatry, 56*(5), 266-272.
- Aldiabat, K. M., & Navenec, C. L. (2011). Philosophical roots of classical grounded theory: Its foundations in symbolic interactionism. *Qualitative Report, 16*(4), 1063-1080.
- American Heritage medical dictionary* (2007). Boston, MA: Houghton Mifflin Co.
- Andersen, J., Øyen, N., Bjorvatn, C., & Gjengedal, E. (2008). Living with long QT syndrome: A qualitative study of coping with increased risk of sudden cardiac death. *Journal of Genetic Counseling, 17*(5), 489-498. doi:10.1007/s10897-008-9167-y
- Auerbach, C. F., & Silverstein, L. B. (2003). *Qualitative data: An introduction to coding and analysis*. New York, NY: New York University Press.
- Barbour, R. S. (2001). Checklists for improving rigour in qualitative research: A case of the tail wagging the dog? *British Medical Journal, 322*, 1115.
<http://dx.doi.org/10.1136/bmj.322.7294.1115>
- Beck, J. S., & Beck, J. S. (2011). *Cognitive behavior therapy: Basics and beyond* (2nd ed.). New York, NY: Guilford Press.
- Bernard, B. (1993). Fostering resiliency in kids. *Educational Leadership, 51*(3), 44-48.
- Bonanno, G. A. (2004). Loss, trauma, and human resilience: Have we underestimated the human capacity to thrive after extremely aversive events? *American Psychologist, 59*(1), 20-28. doi:10.1037/0003-066X.59.1.20

- Burns-Pentecost, A. (2013). *Parents' coping with children's newly diagnosed long-QT syndrome: A qualitative study of the function and utilization of an online user group* (Unpublished doctoral dissertation). Philadelphia College of Osteopathic Medicine, Philadelphia, PA.
- Corbin, J., & Strauss, A. (2008). *Basics of qualitative research: Techniques and procedures for developing grounded theory* (3rd ed.). Los Angeles, CA: Sage Publications.
- CredibleMeds. (2013) A trusted partner providing reliable information on medicines. Retrieved from <http://www.azcert.org/medical-pros/drug-lists/CLQTS.cfm>
- Creswell, J. W., Hanson, W. E., Clark Plano, V. L., & Morales, A. (2007). Qualitative research designs: Selection and implementation. *Counseling Psychologist, 35*(2), 236-264.
- Crotti, L., Celano, G., Dagradi, F., & Schwartz, P. J. (2008). Congenital long QT syndrome. *Orphanet Journal of Rare Diseases, 3*(1), 18. doi:10.1186/1750-1172-3-18
- Dorland's medical dictionary for health consumers.* (2007). Philadelphia, PA: Saunders.
- Earvolino-Ramirez, M. (2007). Resilience: A concept analysis. *Nursing Forum, 42*(2), 73-82. doi:10.1111/j.1744-6198.2007.00070.x
- Eccleston, C., Palermo, T. M., Fisher, E., & Law, E. (2013). Psychological interventions for parents of children and adolescents with chronic illness. *Cochrane Database of Systematic Reviews, 8*, 1-84. doi:10.1002/14651858.CD009660.pub2

- Engle, P. L., Castle, S., & Menon, P. (1996). Child development: Vulnerability and resilience. *Social Science and Medicine*, *43*(5), 621-635. doi:10.1016/0277-9536(96)00110-4
- Etheridge, S. P., Sanatani, S., Cohen, M. I., Albaro, C. A., Saarel, E. V., & Bradley, D. J. (2007). Long QT syndrome in children in the era of implantable defibrillators. *Journal of the American College of Cardiology*, *50*(14), 1335-1340. doi:10.1016/j.jacc.2007.05.042
- Farnsworth, M. M., Fosyth, D., Haglund, C., & Ackerman, M. J. (2006). When I go in to wake them . . . I wonder: Parental perceptions about congenital long QT syndrome. *Journal of the American Academy of Nurse Practitioners*, *18*(6), 284-290. doi:10.1111/j.1745-7599.2006.00132.x
- Gaies, M. G., Watnick, C. S., Gurney, J. G., Bove, E. L., & Goldberg, C. S. (2001). Health-related quality of life in patients with congenitally corrected transposition of the great arteries. *Journal of Thoracic and Cardiovascular Surgery*, *142*(1), 136-141.
- Gallagher, T. (2014). *LQTS: Gender differences and mother-son vs. mother-daughter* (Unpublished doctoral dissertation). Philadelphia College of Osteopathic Medicine, Philadelphia, PA.
- Geelan, D. (2003). *Weaving narrative nets to capture classrooms: Multimethod qualitative approaches for educational research*. Dordrecht, Netherlands: Kluwer Academic.

- Gillespie, B. M., Chaboyer, W., & Wallis, M. (2007). Development of a theoretically derived model of resilience through concept analysis. *Contemporary Nurse, 25*, 124-135.
- Giudicessi, J. R., & Ackerman, M. J. (2013). Genetic testing in heritable cardiac arrhythmia syndromes: Differentiating pathogenic mutations from background genetic noise. *Current Opinion in Cardiology, 28*(1), 63-71.
doi:10.1097/HCO.0b013e32835b0a41
- Giuffre, R. M., Gupta, S., Crawford, S. G., & Leung, A. K. (2008). Fears and anxiety in children with long QT syndrome compared to children with asthma. *Journal of the National Medical Association, 100*(4), 420-424.
- Gomez, R., & McLaren, S. (2006). The association of avoidance coping style, and perceived mother and father support with anxiety/depression among late adolescents: Applicability of resiliency models. *Personality and Individual Differences, 40*(6), 1165-1176. doi:10.1016/j.paid.2005.11.009
- Gonzales, T. (2009). *Confronting the unknown: Mothers' reactions, challenges, and ways of coping with a child's diagnosis of long QT syndrome* (Unpublished doctoral dissertation). Philadelphia College of Osteopathic Medicine, Philadelphia, PA.
- Haynes-Weller, D. (2011). *The experience of grief and loss in persons affected by long QT syndrome* (Unpublished doctoral dissertation). Philadelphia College of Osteopathic Medicine, Philadelphia, PA.
- Herrman, H., Stewart, D. E., Diaz-Granados, N., Berger, E. L., Jackson, B., & Yuen, T. (2011). What is resiliency? *Canadian Journal of Psychiatry, 56*(5), 258-265.

- Jackson, H., Huisman, L., Sanatani, S., & Arbour, L. T. (2011). Long QT syndrome. *Canadian Medical Association Journal, 183*(11), 1272-1275.
doi:10.1503/cmaj.100138
- Janney, A. (2011). *Anxiety in individuals affected by long QT syndrome as experienced by members of an online user group* (Unpublished doctoral dissertation). Philadelphia College of Osteopathic Medicine, Philadelphia, PA.
- Johnson, T. W., & Hanan, P. L. (2001). Long QT syndrome: A prescription for sudden death. *Physician Assistant, 31*-35.
- Kazdin, A. E. (2003). *Research design in clinical psychology* (4th ed.). Boston, MA: Allyn and Bacon.
- Kendall, L., Sloper, P., Lewin, R. J., & Parsons, J. M. (2003). The views of parents concerning the planning of services for rehabilitation of families of children with congenital cardiac disease. *Cardiology in the Young, 13*, 20-27.
- Kendall, L., Sloper, P., Lewin, R. J., & Parsons, J. M. (2003). The views of young people with congenital cardiac disease on designing the services for their treatment. *Cardiology in the Young, 13*, 11-19.
- Knoche, J. W., Orland, K. M., January, C. T., & Maginot, K. R. (2012). Atrial fibrillation and long QT syndrome presenting in a 12-year-old girl. *Case Reports in Pediatrics, 2012*, 124838. doi:10.1155/2012/124838
- Kumpfer, K. L. (1999). Factors and processes contributing to resilience: The resilience framework. In M. D. Glantz & J. L. Johnson (Eds.), *Resilience and development: Positive life adaptations* (pp. 179-224). New York, NY: Kluwer Academic/Plenum.

- Lane, R. D., Reis, H. T., Peterson, D. R., Zareba, W., & Moss, A. J. (2009). Happiness and stress alter susceptibility to cardiac events in long QT syndrome. *Annals of Noninvasive Electrocardiology*, *14*(2), 193-200.
- LaRossa, R. (2005). Grounded theory methods and qualitative family research. *Journal of Marriage and Family*, *67*, 837-857.
- Lazzara, R. (2008). The congenital long QT syndrome: A mask for many faces. *Journal of the American College of Cardiology*, *51*(9), 930-932.
doi:10.1016/j.jacc.2007.11.048
- Liu, J. F., Jons, C., Moss, A. J., McNitt, S., Peterson, D. R., Qi, M., . . . Ackerman, M. J. (2011). Risk factors for recurrent syncope and subsequent fatal or near-fatal events in children and adolescents with long QT syndrome. *Journal of the American College of Cardiology*, *57*(8), 941-950. doi:10.1016/j.jacc.2010.10.025
- Long QT Syndrome. (2012). Retrieved from <http://www.mayoclinic.com/health/long-qt-syndrome/DS00434>
- Long QT Syndrome Support Group. (2013). Retrieved from <http://www.irishheart.ie/iopen24/long-syndrome-support-group-t-221.html>
- Marshall, C., & Rossman, G. B. (2011). *Designing qualitative research* (5th ed.). Thousand Oaks, CA: Sage.
- Martinez, W., Carter, J. S., & Legato, L. J. (2011). Social competence in children with chronic illness: A meta-analytic review. *Journal of Pediatric Psychology*, *36*(8), 878-890. doi:10.1093/jpepsy/jsr035
- Masten, A. S. (2001). Ordinary magic: Resilience processes in development. *American Psychologist*, *56*(3), 227-238. doi:10.1037//0003-066X.56.3.227

- Masten, A. S., Best, K. M., & Garmezy, N. (1990). Resilience and development: Contributions from the study of children who overcome adversity. *Development and Psychopathology*, 2(4), 425-444. doi:10.1017/S0954579400005812
- Masten, A. S., & Coatsworth, J. D. (1998). The development of competence in favorable and unfavorable environments: Lessons from research on successful children. *American Psychologist*, 53(2), 205-220. doi:10.1037//0003-066X.53.2.205
- Mauriello, D. A., Johnson, J. N., & Ackerman, M. J. (2011). Holter monitoring in the evaluation of congenital long QT syndrome. *Pacing and Clinical Electrophysiology*, 34(9), 1100-1104. doi:10.1111/j.1540-8159.2011.03102.x
- Mayo Clinic. (2015). Support groups: Make connections, get help. Retrieved from <http://www.mayoclinic.org/healthy-lifestyle/stress-management/in-depth/support-groups/art-20044655>
- McAdams, D. P., & Olson, B. D. (2010). Personality development: Continuity and change over the life course. *Annual Review of Psychology*, 61(1), 517-542. doi:10.1146/annurev.psych.093008.100507
- Merriam-Webster Online. (2015) Retrieved from <http://www.merriam-webster.com/dictionary/pillar>
- Mosby's Medical Dictionary*. (2009; 8th ed.). Philadelphia, PA: Elsevier.
- Moss, A. J. (2003). Long QT syndrome. *Journal of the American Medical Association*, 289(16), 2041-2044. doi:10.1001/jama.289.16.2041
- Moss, A. J., & Schwartz, P. J. (2005). 25th anniversary of the International Long QT Syndrome Registry: An ongoing quest to uncover the secrets of long QT

syndrome. *Circulation*, *111*(9), 1199-1201.

doi:10.1161/01.CIR.0000157069.91834.DA

Moss, A. J., Schwartz, P. J., Crampton, R. S., Locati, E., & Carleen, E. (1985). The long QT syndrome: A prospective international study. *Circulation*, *1*, 17-21.

doi:10.1161/01.CIR.71.1.17

Nezu, A. M., & Nezu, C. M. (2013). Problem Solving Therapy. Retrieved from <http://www.problemsolvingtherapy.org/index.html>

Nezu, A. M., & Nezu, C. M. (n.d.). Problem Solving Therapy. Retrieved from <http://www.problemsolvingtherapy.org/index.html>

Nezu, A. M., Nezu, C. M., & D’Zurilla, T. J. (2010). Problem-Solving therapy. In N. Kazantzis, M. A. Reinecke, & A. Freeman (Eds.), *Cognitive and behavioral theories in clinical practice* (pp. 76-114). New York, NY: Guilford Press.

Nezu, A. M., Nezu, C. M., & D’Zurilla, T. J. (2013). *Problem-solving therapy: A treatment manual*. New York, NY: Springer.

Nezu, A. M., Nezu, C. M., Felgoise, S. H., McClure, K. S., & Houts, P. S. (2003). Project Genesis: Assessing the efficacy of problem-solving therapy for distressed adult cancer patients. *Journal of Consulting and Clinical Psychology*, *71*(6), 1036-1048. doi:10.1037/0022-006X.71.6.1036

QTSyndrome.ch - History of a heart disease. (2007). Retrieved from <http://www.qtsyndrome.ch/history.html>

Resnick, M. D. (2000). Protective factors, resiliency, and healthy youth development. *Adolescent Medicine*, *11*, 157-165.

- SADS Foundation. (2008). Retrieved from <http://www.sads.org/library/long-qt-syndrome>. VTVUACFViko
- Schwartz, P. J., Moss, A. J., Vincent, G. M., & Crampton, R. S. (1993). Diagnostic criteria for the long QT syndrome: An update. *Circulation*, *88*, 782-784.
doi:10.1161/01.CIR.88.2.782
- Sears, S. F., & Conti, J. B. (2002). Quality of life and psychological functioning of ICD patients. *Heart*, *87*, 488-493.
- Shure, M. B., & Aberson, B. (2006). Enhancing the process of resilience through effective thinking. In S. Goldstein & R. B. Brooks (Eds.), *Handbook of resilience in children* (pp. 373-394). New York, NY: Springer.
- Shure, M. B., & DiGeronimo, T. F. (1994). *Raising a thinking child: Help your young child to resolve everyday conflicts and get along with others*. New York, NY: H. Holt.
- Shure, M. B., & Israeloff, R. (2000). *Raising a thinking preteen: The "I can problem solve" program for 8- to 12-year-olds*. New York, NY: H. Holt.
- Sigelman, C. K., & Rider, E. A. (2012). *Life-span human development* (7th ed.). Belmont, CA: Thomson/Wadsworth.
- Smets, E. M., Stam, M. M., Meulenkamp, T. M., Van Langen, I. M., Wilde, A. A., Wiegman, A., . . . Tibben, A. (2008). Health-related quality of life of children with a positive carrier status for inherited cardiovascular diseases. *American Journal of Medical Genetics Part A*, *146A*, 700-707. doi:10.1002/ajmg.a.32218

- Taggart, N. W., Haglund, C. M., Tester, D. J., & Ackerman, M. J. (2007). Diagnostic miscues in congenital long-QT syndrome. *Circulation, 115*(20), 2613-2620. doi:10.1161/CIRCULATIONAHA.106.661082
- Tusaie, K., & Dyer, J. (2004). Resilience: A historical review of the construct. *Holistic Nursing Practice, 18*, 3-8.
- U.S. Department of Health and Human Services, Administration for Children and Families. (2012). *2012 Resource guide - Preventing child maltreatment and promoting well-being: A network for action*. Retrieved from <https://www.childwelfare.gov/pubpdfs/2012guide.pdf>
- Uzark, K., Jones, K., Slusher, J., Limbers, C. A., Burwinkle, T. M., & Varni, J. W. (2008). Quality of life in children with heart disease as perceived by children and parents. *Pediatrics, 121*(5), E1060-E1067. doi:10.1542/peds.2006-3778
- Varni, J. W. (2013). PedsQL™ (Pediatric Quality of Life Inventory™). Retrieved from http://www.pedsq.org/about_pedsq.html
- Varni, J. W., Limbers, C. A., & Burwinkle, T. M. (2007). Impaired health-related quality of life in children and adolescents with chronic conditions: A comparative analysis of 10 disease clusters and 33 disease categories/severities utilizing the PedsQL™ 4.0 Generic Core Scales. *Health and Quality of Life Outcomes, 5*(1), 43. doi:10.1186/1477-7525-5-43
- Varni, J. W., Seid, M., Knight, T. S., Uzark, K., & Szer, I. S. (2002). The PedsQL 4.0 Generic Core Scales: Sensitivity, responsiveness, and impact on clinical decision making. *Journal of Behavioral Medicine, 25*(2), 175-193.

- Varni, J. W., Seid, M., & Kurtin, P. S. (2001). PedsQL™ 4.0: Reliability and validity of the Pediatric Quality of Life Inventory™ Version 4.0 Generic Core Scales in healthy and patient populations. *Medical Care*, *39*(8), 800-812.
doi:10.1097/00005650-200108000-00006
- Varni, J. W., Sherman, S. A., Burwinkle, T. M., Dickinson, P. E., & Dixon, P. (2004). The PedsQL family impact module: Preliminary reliability and validity. *Health and Quality of Life Outcomes*, *2*(55), 1-6. doi:10.1186/1477-7525-2-55
- Vetter, V. L. (2007). Clues or miscues? How to make the right interpretation and correctly diagnose long QT syndrome. *Circulation*, *115*, 2595-2598.
doi:10.1161/CIRCULATIONAHA.107.700195
- Vincent, G. M. (2005). The long QT and Brugada syndromes: Causes of unexpected syncope and sudden cardiac death in children and young adults. *Seminars in Pediatric Neurology*, *12*(1), 15-24. doi:10.1016/j.spen.2004.11.008
- Wagner, S. M., Lukassen, P., & Mahlendorf, M. (2009). Misused and missed use: Grounded theory and objective hermeneutics as methods for research in industrial marketing. *Industrial Marketing Management*, 1-11.
doi:10.1016/j.indmarman.2008.05.007
- Watson, S. (2011). "Am I going to die today?" *An investigation into the psychosocial impact of long QT Syndrome, healthcare experiences and the genetic testing process* (Unpublished doctoral dissertation). University of Auckland, Auckland, New Zealand.
- Wedekind, H., Burde, D., Zumhagen, S., Debus, V., Burkhardtmaier, G., Mönnig, G., . . . Schulze-Bahr, E. (2009). QT interval prolongation and risk for cardiac

events in genotyped LQTS-index children. *European Journal of Pediatrics*, 168(9), 1107-1115. doi:10.1007/s00431-008-0896-6

Werner, E. E. (2000). Protective factors and individual resilience. In J. P. Shonkoff & S. J. Meisels (Eds.), *Handbook of early childhood intervention* (2nd ed., pp. 115-132). New York, NY: Cambridge University Press. <http://dx.doi.org/10.1017/CBO9780511529320.008>

What is long QT syndrome? (2011). Retrieved from

<http://www.nhlbi.nih.gov/health/health-topics/topics/qt/>

Whitson, S. (2013). Four common defense mechanisms used by stressed out kids.

Retrieved from <https://www.psychologytoday.com/blog/passive-aggressive-diaries/201306/four-common-defense-mechanisms-used-stressed-out-kids>

Zareba, W., Moss, A. J., Locati, E. H., Lehmann, M. H., Peterson, D. R., Hall, W., . . .

Medina, A. (2003). Modulating effects of age and gender on the clinical course of long QT syndrome by genotype. *Journal of the American College of Cardiology*, 42(1), 103-109. doi:10.1016/S0735-1097(03)00554-0

Zolten, K., & Long, N. (2006). *How parents can help their child cope with a chronic illness* (pp. 1-7) (University of Arkansas for Medical Sciences, Department of Pediatrics).

Appendix A
Personal Information Questionnaire

Study ID# _____ ***Date:*** _____

Personal Information Sheet (Primary Caregiver)

Please **fill out** personal information questionnaire for your child diagnosed with LQTS:

Child's Age:

Child Gender:

I.) At what age was your child first diagnosed with LQTS? _____

II.) At what age did your child begin treatment for LQTS? _____

IV) Does your child have any other diagnosed medical or mental health problem?

Yes No

If yes, please describe...

V) Is your child receiving treatment or received treatment in the past for their medical or mental health problem?

Yes No

If yes, please describe...

Information on child with LQTS enrolled in this study

1.) When was your child's first symptomatic episode (i.e., fainting, sudden cardiac arrest)? _____

2.) In what setting did this episode occur?

School Home Sports Other, please specify _____

3.) How old was your child during his/her first LQT event or symptom? _____

4.) When was your child's most recent event and who was present?

Date _____ (month) _____ (year)

Who was present? _____

5. How many events in total (FAINTING, ARREST) has your child had?

0 1 or 2 3 or 4 5 or 6 7 or more

6.) How frequently do your child's LQT symptoms occur?

Never 1-3 times Weekly Monthly Yearly

7.) Has your child witnessed a LQT event (fainting, sudden cardiac arrest) of another family member? Yes No

8.) Does your child have a pacemaker or implantable cardioverter defibrillator (ICD)?

Pacemaker ICD Both Neither

MEDICAL CONSIDERATIONS OF CHILD WITH LQTS

9.) How often does your child take medication?

Once daily 2 or 3 times a day

Other, please specify _____

10.) Who administers medication to your child? Please check all that apply.

Self Parent Teacher School Nurse

Other, please specify _____

11.) Does your child take medication at school? Yes No

12.) Does your child experience side effects from medications? If so, please specify.

Yes _____

No

13.) Does your child know the medication regimen (type of medication, amount, and frequency) he/she is on? Yes No

14.) Does your child follow his/her medication regimen?

Always Almost Always Sometimes Rarely

15.) Is your child involved in the decision-making process and in his/her own therapy and/or treatment? Yes No

16.) Could your child describe what LQTS is to a friend? Yes No

17.) What is the frequency of your child's cardiology appointments?

Every 3 months Every 4 months Twice a year Yearly Other

18.) Does your child have any food restrictions? (i.e. chocolate, grapefruit, licorice, soda, caffeine, other) Yes No

If so, is your child compliant with the restrictions? (i.e. holiday parties, vacations, etc) Yes No

SOCIAL ASPECTS

19.) Has your child been symptomatic in front of their peers? Yes No

If so, under what circumstance(s)? _____

20.) Do any of your child's friends have LQTS? Yes No

Do any of your child's friends have any other chronic disorders? Yes No

If so, what? _____

21.) Does your child require breaks from physical activity for fatigue or rest?

Yes No

22.) What is the estimated number of close friends of your child?

None 1 2 or 3 4 or more

23.) Is your child permitted to sleep over at a friend's house? If so, does he/she?

Yes No

Does he/she spend nights away from home with relatives or others?

Yes No

24.) Is your child involved in any co-curricular activities? (sports, clubs, organizations)

Yes If so, what? _____

No

25.) Is your child restricted from social activities that he/she wants to be involved in?

Yes Please specify _____

No

TEACHERS AND SCHOOL SETTING

26.) What type of school setting is your child enrolled in?

Home schooled Public School Private school

Other_____

27). What is your child's school average?

All A's Mostly A's and B's All B's

Mostly B's and C's_____ All C's _____ Mostly C's and D's

D's and F's

28.) How does your child perform in school relative to others their age?

Reading: Above average Somewhat above average Average

Below Average Somewhat below average Below average

Math: Above average Somewhat above average Average

Below Average Somewhat below average Below average

29.) Does your child receive accommodations through the school district? If so, what are they?

Yes Please Specify_____

No

30.) Does your child have any problems within school, such as school refusal, academic difficulty, or peer group? If so, please specify. Yes No

31.) Are your child's teachers aware that he/she has LQTS? Yes No

32.) Has your child moved or changed school districts? If so, how many times? Why?

Yes

How many times? _____

Reason? _____

No

**INFORMATION REQUESTED ABOUT PRIMARY CAREGIVER AND/OR
SPOUSE/PARTNER**

33.) Age of Primary Caregiver _____

What is your relationship to the child with LQTS? _____

Diagnosed with LQTS? Yes No

If so, symptomatic? Yes No

34.) Age of spouse/partner _____

Diagnosed with LQTS? Yes No

If so, symptomatic? Yes No

What is the spouse/partner's relationship to the child with LQTS? _____

Is the spouse/partner related to the child? Yes, If so, how? _____ No

THANK YOU VERY MUCH FOR YOUR PARTICIPATION!

Appendix B

Interview Questions

Introduction with Parent(s)

Thank you for taking the time to meet with me and allowing your child to participate in my study. The purpose of my study is to learn what it is like for a child to live with Long QT Syndrome from the child's perspective. My hope is that the information we learn will help others to better understand what a child thinks and feels about Long QT Syndrome. I have a few questions to ask you before I start the interview.

Discussion with Parent(s)

1. Are there any significant changes that have occurred in the past year for your child in their home, school, or well-being?
2. How do you refer to Long QT or your child's heart condition when you speak to your child?
3. Is it ok for me to ask your child what he/she knows about Long QT?
4. Do you have any questions before I start the interview with your child? (If yes, answer any questions. If no, meet with child to start interview).

PedsQL

1. Completion of the PedsQL (Parent version).

Introduction with Children

Thank you for taking time to meet with me! . I would like to ask you some questions about how you think and feel, and about things you do, if at any time you want to take a

break please let me know. Do you have any questions before we start? (If yes, answer any questions. If no, state, “Ok, Let’s begin”).

Interview with Children

Get to Know You

1. If you were able to have a special/magical power, what would it be?
2. If you were given three wishes? What would they be?
3. Where do you go to school?
4. What do you like about school?
5. What do you dislike about school?
6. If you could change anything about school, what would it be?
7. Tell me about a good friend of yours.
8. Have you ever slept at a friend’s house overnight? (If yes, how was it? If no, would you want to?)
9. What do you do for fun?
10. Do you play any sports? (If yes, what are they? If no, why?)
11. What are your favorite things to do outside/outdoors?

Understanding of LQTS (Knowledge)

1. Why do you go to a heart doctor?

- 2 *What do you know about LQTS? (based on first question)
- 3 How do you feel about going to the doctor?
- 4 If you had a friend who had to go see your heart doctor, what would say to them before they go?
- 5 When you know you have to go to see the heart doctor, do you talk with your friends about it? *(What do you tell them?)
- 6 What do you have to do to keep your heart healthy? (Or is there anything special you have to do for your heart?)
- 7 Do you ever wear a heart monitor? If someone asks you why you wear a heart monitor, what do you say?
- 8 Do your friends ask you about why you need to take medication?
- 9 *Do people in school know that you have Long QT? *Do you think they know what it is? What do you tell them? How do you feel about that?
- 10 Do you take medicine at school or see the school nurse frequently? What do you think about that?
- 11 Is there anything you are not allowed to do at school that other kids are allowed to do? (Prompt: Gym, Lunch, Recess)
- 12 Does your family worry about your heart (or keeping your heart healthy)? Who in your family worries about you? *(How do you know, or what do they say?)

13 *How does that make you feel?

14 Does your parent/s treat you and your brother/s or sister/s the same? *(Tell me about it.)

15 What ways are different?

16 Is there anything your parent/s doesn't let you do because of your heart that you wish they would?

17 Is there anything that you want to tell me that I forgot to ask or that I should know about you or Long QTS?

18 What did you think about this meeting?

19 Do you have any questions for me? Do you want to know my favorite super power that I wish I had?

Conclusion

Thank you so much for meeting with me! I really liked getting to know you.

PedsQL

1. Completion of the PedsQL generic core scale and the cardiac module (age appropriate)