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Siblings of Children with Long QT Syndrome: Relationships and Coping

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Philadelphia College of Osteopathic Medicine

Department of Psychology

SIBLINGS OF CHILDREN WITH LONG QT SYNDROME: RELATIONSHIPS AND
COPING

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Submitted in Partial Fulfillment of the Requirements for the Degree of

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DEPARTMENT OF PSYCHOLOGY

Dissertation Approval

This is to certify that the thesis presented to us by Courtney Melven-Wurst
on the 19th day of August, 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.

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Dedication

This dissertation is dedicated to the unaffected siblings of children with LQTS who volunteered their time to participate in this study. It is because of your openness to share your experiences about the effects of chronic illness on your families that other siblings may benefit in the future. This is also dedicated to my sisters, who were the inspiration for this study, as they knew what it was like to live with a sibling with chronic illness. In particular, this is dedicated to my sister, Megan Fitzgibbon, who was my ultimate protector, often watching me while I slept to make sure I was okay. Lastly, this dissertation is dedicated to the memory of my mother, Donna McCuen, who left this earth way too soon.

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Above all others, I would like to express my love and gratitude for my husband Kevin and my son, Lucas. There are no words to express how much your love, support, and confidence has meant to me throughout this program. Thank you, Kevin, for the countless back rubs and words of encouragement during those late nights writing papers, particularly this dissertation. Thank you, Lucas, for being my self-care and helping maintain my sanity with your laughter and love. Last, but certainly not least, I would like to thank my parents who never lost faith that I would succeed. Without your love, admiration, guidance and support, I would not be the person I am today. Thank you.

Abstract

Long QT syndrome (LQTS) is a congenital heart disorder, usually diagnosed in childhood, that may lead to cardiac arrest, seizures, syncope, and sudden death. The birth of a child with LQTS, as well as the social, physical, and psychological functioning of the affected child, can have a significant impact on the family. The family stress associated with having a child with any disability or chronic illness is likely to affect the lives of siblings. The lack of research on the effects of having a sibling with long QT syndrome on individuals who do not have LQTS provided the rationale for the current study. A qualitative research design was utilized to determine if and how LQTS impacts quality of life of unaffected siblings of children with long QT syndrome. Two themes emerged: relationships and coping strategies. A majority of the participants maintained positive relationships with their parents and affected siblings and had friends/support systems both within and outside of the home. Participants coped with the diagnosis and management of their siblings' LQTS by trusting the medical team and obtaining information about the illness. A majority of the participants did not indicate that their lives differed greatly from siblings in other families not affected by LQTS.

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Chapter 1: Introduction

Statement of the Problem

In the United States, it is estimated that 5% to 31% of children experience moderate to severe chronic illness or disability (Cox, Marshall, Mandelco, & Olsen, 2003). *Chronic illness* refers to an ailment that persists for 6 months or more and requires continuing medical treatment, permanent lifestyle changes, and constant adaptation to an unpredictable course (Perrin et al., 1993). Chronic illnesses, such as asthma, sickle cell anemia, and epilepsy, have no known cures, but can be managed medically (Perrin et al., 1993). As indicated by Newacheck and Taylor (1992), chronic illness has changed considerably in the last century in that there have been significant improvements in management of symptoms, allowing those affected to live longer. For example, studies have shown a sevenfold increase in survival until the age of 21 in children with cystic fibrosis and a twofold or more increase in survival rates of children with spina bifida and congenital heart disease (Gortmaker, 1985). Long QT syndrome is a congenital heart disorder, usually diagnosed in childhood, that may lead to cardiac arrest, seizures, syncope, and sudden death (Garson et al., 1993). It occurs in approximately 1 in every 2,000 live births and is characterized by an abnormal QT-interval prolongation and an increased risk of death, usually due to ventricular fibrillation (Roden, 2008; Schwartz et al., 2010). The disorder affects both children and young adults with structurally normal hearts (Moss et al., 1991). Syncope and sudden death associated with long QT syndrome can be triggered by physical and emotional stressors. At times, these events can also be triggered by a loud noise or occur when the individual

is resting (Roden, 2008). Risk is influenced by gender, age, genotype, environmental factors, and therapy (Goldenberg, Zareba, & Moss, 2008). Due to the unpredictable nature of chronic illnesses, the birth of a child with a chronic illness, as well as the social, physical, and psychological functioning of the affected child, can have a significant impact on the family.

The family systems perspective recognizes that each member of the family influences the others. As a result, the way in which parents cope with chronic illness can affect the child and other members of the family. Family systems theory is derived from systems theory, which views any system as a whole, with elements that are dependent on one another (Minuchin, 1985). The family is a system in that a change in the functioning of one family member is followed by a compensatory change in another member of the family (Bowen, 1974). Based on this perspective, the family stress associated with having a child with any disability or chronic illness is likely to affect the lives of siblings (Cox et al., 2003).

Because the sibling relationship is among the most influential and long lasting of human relationships (Gallagher, Powell, & Rhodes, 2006), the health and functioning of one child may affect the health and functioning of a sibling (Cox et al., 2003). Studies on the impact of chronic illness on siblings have shown both positive and negative outcomes regarding cognitive, social, and psychological functioning (Derouin & Jessee, 1996).

As indicated by Cox et al. (2003), most research on siblings of children with disabilities suggests one or more negative effects. The most commonly cited negative effects on siblings of children with chronic illness and disabilities are maladjustment and behavioral problems (Cox et al., 2003). Behavioral and emotional problems include

struggles in school, jealousy, decreased self-esteem, increased impulsivity, and depressive affect (Cox et al., 2003). In addition to behavioral and emotional issues, siblings of children with disabilities may experience an increase in responsibilities, including household chores and child care (Cox et al., 2003). Siblings may also become a source of comfort and support for parents struggling with their child's disability (Cox et al., 2003). Both consequences may lead to parentification of siblings (Lamorey, 1999). In short, having a sibling with a disability or chronic illness may negatively impact quality of life.

According to Bonomi, Patrick, Bushnell, and Martin (2000), the World Health Organization indicates that quality of life is a subjective concept that focuses on the self-perception of one's current state. Quality of life includes health status, psychological well-being, and social functioning (Bonomi et al., 2000). In addition, quality of life incorporates level of independence, personal beliefs, and relationships to features of the environment (Bonomi et al., 2000). Wrosch and Scheier (2003) state that a good quality of life indicates successful growth as a person. A good quality of life also indicates that the hopes of an individual have been matched by his or her experiences. A poor quality of life, however, occurs when an individual's hopes do not match his or her experiences (Skevington, O'Connell, & the WHOQOL Group, 2004).

Quality of life, as defined by the World Health Organization, may be positively or negatively influenced by the birth of a child with a chronic illness. If siblings of children with chronic illness or disabilities are vulnerable to poor psychosocial outcomes (Breslau, Weitzman, & Messenger, 1981; Lavigne & Ryan, 1979), then it is important to address

the factors that may contribute to these outcomes in order to increase the chances for a good quality of life (Bellin & Kovacs, 2006).

Purpose of the Study

Although there is a great deal of research examining the social, physical, and psychological adjustment of both the family and siblings of children with disabilities and other chronic medical illnesses, no available research has focused solely on the functioning of siblings of children with long QT syndrome. Most research on children with LQTS has focused on the child or on the impact on the family as a whole without specific attention given to the siblings. The aim of the present study was to learn how LQTS impacts unaffected siblings of children with LQTS.

Literature Review

Chronic illness.

Chronic illness is a common phenomenon that affects many families. In addition to requiring continuous medical attention and permanent lifestyle changes (Kyngas, Kroll, & Duffy, 2000), chronic illness is also characterized by an erratic course with periods of acute crises (LeBlanc, Goldsmith, & Patel, 2003). Due to advances in pediatric medicine, there has been an increase in the number of chronically ill children surviving into adulthood (Halfon & Newacheck, 2010; Newacheck & Taylor, 1992). As a result, chronically ill children may require support from their families, including siblings, throughout their lives.

Long QT syndrome.

Long QT syndrome is a congenital disorder, usually diagnosed in childhood, in which mutations affecting the cardiac ion channels result in delayed ventricular repolarization

and a prolonged QT interval (Liu et al., 2011). The associated arrhythmias may be marked by syncope, aborted cardiac arrest, or sudden cardiac death (SCD) (Liu et al., 2011). Although the disorder can be fatal, it is highly treatable (Giudicessi & Ackerman, 2013). The syndrome is considered to be responsible for 2,000 to 3,000 unexpected deaths in children and adolescents each year (Modell, Bradley, & Lehmann, 2012; Perez, Kumarasamy, Owens, Wang, & Hlatky, 2011). Research suggests that the disorder is caused by mutations in genes that encode the structure of cardiac ion channels (Zareba et al., 2003). Syncope and sudden death associated with long QT syndrome can be triggered by physical and emotional stressors, as well as by a loud noise or by resting, depending on the syndrome subtype (Roden, 2008). Long QT syndrome is “probably the most important life-threatening arrhythmogenic ion channel disease” affecting young people (Schwartz et al., 2010, p. 1273).

Most affected individuals have delayed ventricular repolarization evident on the electrocardiogram (ECG), a recording of the electric activity of the heart (Goldenberg et al., 2008). Long QT syndrome occurs in response to mutations involving the myocyte ion channels and has an autosomal-dominant inheritance pattern (Goldenberg et al., 2008). Over the past 20 years, 15 LQTS susceptibility genes have been identified using classical linkage analysis (Giudicessi & Ackerman, 2013). More than 500 mutations have been identified in 12 LQTS- susceptibility genes, with the type 1 (LQT1), type 2 (LQT2), and type 3 (LQT3) genotypes comprising more than 95% of genotype-positive LQTS and approximately 75% of all LQTS (Goldenberg & Moss, 2008). Approximately 85% of reported cases inherit the gene from one parent; the remaining 15% have new mutations that are not inherited (Goldenberg et al., 2008).

However, in recent years, researchers have discovered that LQTS, although genetic, is subject to incomplete penetrance, as well as variability in the ways it may be expressed. For example, genotype-positive individuals may have a variety of clinical phenotypes, ranging from a continuous asymptomatic state to sudden death in infancy. Approximately 25% of genotype-positive LQTS patients do not have any overt symptoms (Giudicessi & Ackerman, 2013). Additional research has contributed to the understanding that a combination of genetic and environmental factors modify symptom onset, degree of QTc prolongation, and risk of LQTS-triggered cardiac events (Giudicessi & Ackerman, 2013).

Subtypes of long QT syndrome.

Research suggests that there are 13 subtypes of LQTS (Gonzalez, Veneziano, Puggina, & Boccia, 2015). LQT1 is the most common form of the disease and occurs from loss-of-function mutations in KCNQ1, which predetermines an adrenergic-sensitive potassium current in the heart (Roden, 2008). In LQT1 cases, syncope or sudden death is triggered by physical or emotional stressors. For example, swimming and diving are triggers for stress in this population. LQT2 occurs from loss-of-function mutations in KCNH2, which predetermines another important potassium current in the heart. In these cases, syncope or sudden death can occur from stress or periods of rest (Schwartz et al., 2001). Triggering events may include sudden loud noises, such as an alarm clock (Roden, 2008). The LQT3 subtype occurs from mutations that interrupt fast inactivation of the cardiac sodium-channel SCN5A. As a result, the sodium current persists abnormally during the cardiac action potential, prolonging the QT interval (Roden, 2008).

Similar to LQT2, events in LQT3 cases, such as syncope and sudden death, occur during periods of rest and sleep (Anderson, Øyen, Bjorvatn, & Gjengedal, 2008).

Risk factors related to age and gender.

As indicated by Locati et al. (1998), a female predominance in congenital long QT syndrome has been reported, but not fully explained. In an initial report on the International LQTS Registry in which 186 individuals were enrolled, females had a higher risk of cardiac events than did males (Moss et al., 1985). An increased female prevalence was consistently found among the patients referred to the LQTS registry (Moss et al., 1991).

In order to determine the effects of sex and age on LQTS genotypes, Zareba et al. (2003) collected data on and followed 533 patients: 243 LQT1 gene carriers, 209 LQT2 gene carriers, and 81 LQT3 gene carriers. The probability of cardiac events was analyzed by gender, age, and genotype. Researchers found that during childhood, the risk of cardiac events, including syncope, aborted cardiac arrest, or sudden death, was significantly higher in LQT1 males than in females with the LQT1 genotype (Zareba et al., 2003). In childhood, researchers found no significant differences between males and females with the LQT2 and LQT3 genotypes. As adults, LQT2 and LQT1 females had a significantly higher risk of cardiac events than their male counterparts (Zareba et al., 2003).

The diagnosis of long QT syndrome is based primarily on QT interval duration (Roden, 2008). QT interval, however, is often age- and sex-dependent. For example, in a community-based study conducted by Rautaharju et al. (1992), the researchers attempted to determine if sex differences were evident in the evolution of the QT interval.

They found that the QT interval was significantly longer in females in all age groups from 15 to 50 years of age. Upon further investigation, the researchers concluded that the sex difference was explained by QT interval shortening in males after puberty, as opposed to a prolongation in females during reproductive years (Rautaharju et al., 1992). A subsequent study concluded that sex differences in QT interval are not present at birth, indicating that the vulnerability to cardiac events changes with age for LQTS males and remains increased for LQTS females (Stramba-Badiale, Spagnolo, Bosi, & Schwartz, 1995; Zareba et al., 2003).

Diagnosing long QT syndrome.

T-wave morphology in an ECG may be useful in diagnosing LQTS. Moss et al. (1995) described different patterns of repolarization morphology in LQTS, including flat T-waves, broad-based T-waves with a slow rise of the initial segment, peaked T-waves, and notched T-waves. Moss et al. (1995) evaluated the relationship between T-wave morphology and genotype and found that LQT1 was characterized by wide, broad-based T-waves, while LQT2 was characterized by low amplitude and frequently notched T-waves. LQT3 was characterized by a long ST segment, followed by a peaked, tall T-wave.

Roden (2008) notes that the most common LQTS presentations are palpitations, presyncope, syncope, and cardiac arrest. Asymptomatic individuals may be evaluated for LQTS if a family member has been diagnosed with the syndrome. Differential diagnoses can vary from less serious conditions, such as vasovagal syncope, to more serious conditions, such as hypertrophic cardiomyopathy (Roden, 2008). An abnormal ECG acquired while the patient is at rest is vital in diagnosing the syndrome (Roden, 2008).

The QT interval noted during the ECG is affected by heart rate and gender. The upper limits of the QT interval corrected for the heart rate are below 460 milliseconds for women and below 440 milliseconds for men. However, normal QTc values do not exclude a diagnosis of LQTS. An estimated 10% to 40% of genotype-positive individuals do not have any evidence of a QT abnormality and are categorized as “normal QT interval” LQTS or “concealed” LQTS (Priori, Napolitano, & Schwartz, 1999; Tester, Will, Haglund, & Ackerman, 2006). In addition to QT interval and symptoms characteristic of LQTS, a detailed family and personal history can also contribute to diagnosing the syndrome, especially in cases of concealed LQTS (Roden, 2008). The evaluator should ask not only about sudden deaths in the family, but also about deaths due to drowning, driving, or sudden infant death syndrome (SIDS), all of which can be indicators of LQTS (Roden, 2008).

Treatment of long QT syndrome.

Individuals with a low risk of sudden death do not require treatment, although it is important to avoid drugs that can prolong the QT interval (Roden, 2008). In addition, and regardless of symptom status, individuals with LQTS should maintain adequate hydration (Giudicessi & Ackerman, 2013). The first line of therapy in treating LQTS is beta blockers (Goldenberg et al., 2008). Beta blockers should be administered in people at intermediate and high risk; beta blockers should be considered on an individual basis for low-risk cases (Goldenberg et al., 2008). In a study conducted by the International LQTS Registry, beta blockers were shown to reduce the frequency of syncope, although they have little or no effect on QTc duration (Moss et al., 2000). The reduction in syncope episodes was more evident in high-risk patients who experienced the highest

event rates prior to beginning beta-blocker therapy. It is important to note that beta-blocker therapy did not reduce cardiac events in individuals with LQT3 mutations (Goldenberg et al., 2008). Research has also suggested that beta-blocker therapy decreased the likelihood of syncope or sudden death among patients with the LQT1 subtype more than with patients with the LQT2 or LQT3 subtypes (Priori et al., 2004). As indicated by Schwartz, Crotti, and Insolia (2012), beta-blocker therapy should be initiated in all patients, even those who remain asymptomatic, as 10%–12% of LQTS cases first manifest as sudden death.

Beta-blocker therapy in addition to an implantable cardioverter defibrillator (ICD) is considered the second line of prevention in individuals with LQTS (Goldenberg et al., 2008). The combination therapy is indicated as the primary prevention method for high-risk individuals who continue to experience events despite beta-blocker therapy (Goldenberg et al., 2008). The ICD is an effective form of both primary and secondary prevention in individuals with LQTS (Groh et al., 1996; Zareba et al., 2003). Patients with LQTS who receive an ICD are usually younger than recipients with acquired cardiac diseases; therefore, patients with LQTS may be exposed to the negative consequences of receiving an ICD (Goldenberg et al., 2008). Complications of the device include pain of shocks, lead-related complications, infection, and psychological adjustment to the device (Goldenberg et al., 2008). In some cases, particularly in malignant forms of LQTS or when beta blockers have been unsuccessful, a surgical intervention referred to as left cardiac sympathetic denervation, which involves the removal of thoracic ganglia, may be necessary (Schwartz et al., 2012).

Family systems theory.

Systems theory focuses on the functioning of a system and of its individual components. A derivative of systems theory, family systems theory focuses on the family as a whole, as well as each member of the family unit (Bowen, 1974). Gallagher, Powell, and Rhodes (2006) note the family can be viewed as a unified system that supports the interdependence of each member. Interactions between various members of the family can influence the interactions between other members of the family. In addition, a change in one family member can lead to changes in other members of the family unit (Gallagher et al., 2006).

When the functioning of one family member changes, another family member automatically compensates for that change (Bowen, 1974). For example, when one member of the family becomes ill, another member or other members will compensate for the change by overfunctioning within the family. If the sick family member becomes chronically ill, the overfunctioning of other family members may create a long-term imbalance in the family. Systems theory also suggests that evolution and change are inherent; the family must adapt to change in order to maintain homeostasis. In other words, when the pattern of the family is disturbed, such as when a child is diagnosed with a chronic illness, the family must challenge the existing pattern, explore alternatives, and embrace emergent patterns that are more appropriate to the changed circumstances (Minuchin, 1985).

Systems theory also suggests that complex systems have subsystems. Each family is composed of subsystems, including the spouse subsystem, the parent subsystem, the parent-child subsystem, and the sibling subsystem (Minuchin, 1985). Systems theory

further suggests that each subsystem is separated by boundaries, and interactions across those boundaries are determined by rules and patterns (Minuchin, 1985). Again, if homeostasis is disrupted when a child is diagnosed with a chronic illness, the rules and boundaries regarding interactions between the subsystems must change. In sum, the diagnosis of a chronic illness in a family member, particularly a child, can significantly impact the family as a whole.

Effects of chronic illness and disability on family.

The diagnosis and management of a chronic illness in a family member, particularly a child, creates both short- and long-term stressors for the entire family (LeBlanc et al., 2003). It may also impact the quality of life of the family as a whole. Family quality of life considers all family members in terms of what is required to have a good life as a whole unit (Poston, Park, Turnbull, Mannan, & Marquis, 2003). The impact of a disease and treatment on family functioning is a concern, given the role of the family in the child's adaptation to the illness (Varni et al., 2004). Family members influence one another both directly and indirectly. Parents and siblings can influence each other directly. For example, if the mother and/or father of the affected child can not adapt to the chronic illness, a sibling would have difficulty adapting. Indirectly, each member of the family, through his or her relationship with another family member, impacts another member of the family (Parke, 2004). Varni et al. (2004) conducted a study in which parents of 23 pediatric patients with chronic illness and 12 parents of pediatric patients living in a long-term facility completed the PedsQLTM Family Impact Module. The 5-point scale measured responses to questions about physical, emotional, social, and cognitive functioning and about communication, worry, daily activities, and

family relationships. The parents with ill children living at home reported lower functioning in terms of family relationships, communication, and emotional functioning (Varni et al., 2004).

Some emotions experienced by parents and family members of affected children may include grief and feelings of loss. Grief is often associated with the diagnosis of a chronic illness in childhood (Worthington, 1989). One major cause of initial grief is the idea that parents grieve the loss of the “perfect” child they had envisioned. The next cause of grief for parents centers on the child they have now. This grief includes concerns about the child’s quality of life and uncertain life expectancy (Worthington, 1989). A third source of grief focuses on the impact that the chronic illness will have on the family as a whole. Parents are concerned about the impact of the illness on the demands of daily life (Worthington, 1989). Impacts on the family include financial, social, psychological, and overall family functioning (LeBlanc et al., 2003).

Parents of children with a chronic illness, such as long QT syndrome, may experience financial challenges in the form of inability to work, frequent visits to physicians or the hospital, and the costs associated with medical regimens for the ill child (Breslau, Staruch, & Mortimer, 1982; LeBlanc et al., 2003). In addition to experiencing economic challenges, families with children suffering from chronic illness also experience social and psychological impacts. Social effects of chronic illness are greater for mothers, as they are usually the primary caretakers (LeBlanc et al., 2003). Mothers of children with chronic illness may experience negative social effects, including decreased social contacts and activities outside of the home (LeBlanc et al., 2003). For example, children with the LQTS1 genotype are often restricted in the types of extracurricular

activities they are allowed to participate in, including organized sports (Roden, 2008). As a result, the primary caregiver may also be restricted in the number of activities they attend or participate in outside of the home, possibly leading to a sense of isolation.

Consistent with family systems theory, family structure may need to be adjusted and roles may be compartmentalized when a child is diagnosed with a chronic illness (LeBlanc et al., 2003). After a child is diagnosed, a previously employed mother may become the primary caretaker, while the father functions as the sole breadwinner, or vice versa. The progression of the caretaker-ill child relationship may become enmeshed and overprotective, while the economic provider becomes distant from the child and other members of the family (LeBlanc et al., 2003). In a study conducted by Breslau, Staruch, and Mortimer (1982), two indexes of psychological distress in mothers of children with cystic fibrosis, cerebral palsy, or multiple physical handicaps were examined; there was also a control group. These indexes measured distress based on a depression-anxiety scale and a 7-item scale created by Pearlin and Schooler (1978) measuring “unpleasant feelings of which people are aware” (e.g., frustration, tension, worry) specific to a woman’s experience as a mother (Breslau et al., 1982, p. 683). The researchers found that mothers of children with disabilities scored significantly higher than the control group on both indexes. In addition, the more dependent the disabled child, the greater the mother’s distress (Breslau et al., 1982).

Although there can be many challenges when a child is diagnosed with a chronic illness or disability, studies have shown that the experience can also have a positive impact on the family. In a study conducted by Taunt and Hastings (2002), families of children with disabilities were recruited. The most frequently occurring disabilities were

Down syndrome, Asperger syndrome, and cerebral palsy. One group was interviewed in person, while the other was given an online survey with open-ended questions. Both groups were asked what they believed to be positive aspects of caring for their child, whether they believed the presence of the disabled child had positive effects on their other children and extended family, and what they believed to be particularly rewarding about the experience. The researchers found significant overlap in both groups concerning positive aspects, including increased sensitivity, opportunities to learn, and changes on life perspective. Additional positive aspects included opportunities to expand one's social network and increased assertiveness (Taunt & Hastings, 2002).

Effects of chronic illness and disability on siblings.

Although the research is limited, a few studies have focused solely on the siblings of children with chronic illness and disabilities. Based on the family systems perspective, the family stress associated with having a child with any disability or chronic illness is likely to affect the lives of siblings (Cox et al., 2003). Observational and interview studies support the idea that children may respond emotionally or behaviorally to a sibling's illness (Lavigne & Ryan, 1979). In a study conducted by Lavigne and Ryan (1979), siblings of chronically ill children, ages 3 to 13 years, were compared to siblings of healthy children in order to examine psychological adjustment. The adjustment measure used was a paper and pencil, objective measure of children's emotional and behavioral issues. The researchers found that siblings of children with chronic illness had more symptoms of irritability and social withdrawal. In addition, differences in chronic illness (hematology, plastic surgery, and cardiology patients) neared significance on measures of fear and inhibition (Lavigne & Ryan, 1979).

Although long QT syndrome is not a disability, but rather a chronic illness, it is helpful to look at the effects of both on siblings because it can affect the family in similar ways. As noted above, maladjustment and behavioral problems are the most common more negative effects (Cox et al., 2003). In a study conducted by Breslau, Weitzman, and Messenger (1981), mothers completed a psychiatric screening inventory for children between the ages of 6 and 18 years old with or without disabled siblings. The inventory was administered during a 2- to 3-hour visit in the home; it measured the unaffected siblings' level of functioning with parents, siblings, peers, and teachers. The researchers found that the siblings of disabled children scored significantly higher in mentation problems, fighting, and delinquency.

Not all effects on siblings of children with disabilities are negative; research has also identified positive consequences of having a sibling with a disability. Coping responses among siblings of children with disabilities can determine overall adjustment. The study conducted by Cox et al. (2003) found that siblings of children with disabilities have increased empathy for others and are less self-centered. Cox et al. (2003) conducted a qualitative study to determine coping responses to daily life stressors of siblings of children with disabilities. Siblings were between the ages of 6 and 18 years. Disabilities in the sample included mental retardation, Down syndrome, autism, and sight and hearing impairments. The sibling older than and closest in age to the disabled child was asked to complete an open-ended sentence-completion task involving stressful situations (Cox et al., 2003). Coping reactions that emerged included proactive, nonactive, interactive, and internally reactive. The researchers found that most responses were proactive; siblings took responsibility for improving a stressful situation using problem-solving skills. The

second most common reaction was interactive; siblings sought social support to manage the situation. The third most common response was internally reactive; siblings responded to a situation by expressing emotions or thinking about things more carefully. The least common reaction was nonactive; siblings in this group did not demonstrate a physical or emotional response to a stressful situation. The researchers indicated that these findings may support other research that suggests siblings of disabled children have increased responsibility in the family, hence the most common response to be proactive (Cox et al., 2003).

Siblings of children with chronic illnesses may also be at risk of psychological maladjustment. According to Derouin and Jessee (1996), dealing with chronically ill children can have a profound effect on all members of the family. Similar to studies of siblings of disabled children, studies on the impact of chronic illness on siblings have shown both positive and negative outcomes in psychological functioning (Derouin & Jessee, 1996). The authors suggest that the range of outcomes and discrepancies may be based on specific diagnoses and maternal reports of sibling adjustment (Derouin & Jessee, 1996). For example, the impact of LQTS on siblings may change, depending on whether the child has the LQT1, LQT2, or LQT3 genotype, as restrictions and treatment can vary. In their study, Derouin and Jessee (1996) aimed to determine perceptions of family disruption and self-esteem in siblings of children with asthma or cystic fibrosis. Fifteen families participated; there were 6 male and 9 female participants, with a mean age of 10.1 years, with the ill sibling being, on average, younger than the participants. All of the parents were married and had an average of two or three children. A control group of siblings of healthy children was also used in the study. Siblings in the healthy

group were matched by sex and age to siblings in the asthma and cystic fibrosis groups. Siblings completed a semistructured phone interview with the investigator (Derouin & Jessee, 1996). The investigator asked questions related to knowledge of the illness, impact on the ill child, effect of illness on the family, and effect of illness on the responding sibling. The researchers found that a majority of the siblings noted changes in the chronically ill child physically and behaviorally. Siblings in the asthma group noted that the ill child was more playful, while siblings in the cystic fibrosis group noted that the ill child had gained weight (Derouin & Jessee, 1996). Over half of the siblings in either group did not believe the ill child received special attention, but 42% believed that their ill sibling did receive special attention (Derouin & Jessee, 1996). Eighty-nine percent of siblings perceived their parents as being worried about the health of the chronically ill child (Derouin & Jessee, 1996). Siblings were able to identify both positive and negative effects of having a chronically ill brother or sister. Siblings believed that positive outcomes included strengthening of the family and being involved in special events for the chronically ill child (Derouin & Jessee, 1996). Worrying about the ill child and the disruption of everyday activities were the most common negative effects reported by the siblings (Derouin & Jessee, 1996). Overall, 40% of the siblings in the cystic fibrosis group and 11% of the siblings in the asthma group reported being the most unhappy member of the family due to the child's illness (Derouin & Jessee, 1996).

As indicated by Rana and Mishra (2015), having a sibling with a chronic illness, specifically a neurological disorder, can result in the unaffected sibling feeling stress associated with embarrassment around peers, trying to make up for the deficits of the affected sibling, and concern regarding parental stress and grief. The researchers' study

consisted of 50 siblings, ages 12 to 18 years old, of children with chronic neurological disorders, including epilepsy, cerebral palsy, autism spectrum disorders, and mental retardation. A control group of sex- and age-matched peers was also included in the study. The participants completed a personal history questionnaire, as well as a quality of life measure, specifically focused on physical, social, psychological, and environmental factors. A majority of the children were not aware of the nature of the affected siblings' disorder. The researchers found that 74% of the control group and 40% of the children in the study group had adequate energy for daily activities. Those in the study group needed more medical treatment to function in their daily life than the control group. Forty-four percent of children in the study group were not satisfied with their capacity to work, and 14% of the children reported being dissatisfied or very dissatisfied with their sleep. In the area of psychological factors, 68% of the study group reported enjoying life, compared to 90% of the control group. Fifty-four percent of the children in the study group reported an inability to concentrate fully on their work and also reported little opportunity for leisure activities. Overall, the authors suggest that the quality of life of unaffected siblings is significantly negatively impacted in adolescents (Rana & Mishra, 2015).

Although there have been many studies that found negative effects on the unaffected siblings of children with chronic illness, others studies have demonstrated positive effects, including increased empathy and an ability or willingness to take on more responsibility (Havermans, De Croock, Vercruysse, Goethals, & Van Diest, 2015). In a study conducted by Havermans et al. (2015), siblings of children with varying illnesses, including congenital heart disease, cystic fibrosis, cancer, and type 1 diabetes,

were recruited to complete questionnaires measuring quality of life. The participants ranged in age from 10 to 18 years old. There was a sex- and age-matched control group from a previous study. The researchers had four hypotheses: that the unaffected siblings would rate their quality of life lower than controls, that younger siblings would report a better quality of life than older siblings, that female siblings would report a lower quality of life than males, and a longer time since diagnosis would be related to a higher quality of life and lower impact of illness. The researchers found no significant differences between the study and control groups. Contrary to previous literature, siblings reported a good quality of life, similar to siblings of healthy children.

Houtzager, Grootenhuis, Hoekstra-Weebers, and Last (2005) indicate that psychosocial problems that are present in siblings prior to the onset or diagnosis of an illness may be a sign of vulnerability to adjustment problems. Other researchers have stated that the illness of a family member not only causes new illness related problems, but that preexisting psychological and social issues may be exacerbated in many cases (Kalnins, Churchill, & Terry, 1980). In a study conducted by Sahler et al. (1994), the emotional and behavioral distress in siblings of children with cancer was assessed. Parents and siblings were interviewed and completed self-report measures; a matched control group was included, as well. The study found that 40% of children with emotional or behavioral problems prior to the diagnosis of cancer in a sibling had more psychosocial adjustment problems after the diagnosis.

Siblings of children with chronic or life-threatening illnesses may experience internalized problems, including anxiety and feelings of isolation (Houtzager, Oort, Hoekstra-Weebers, Caron, Grootenhuis, & Last, 2004). Houtzager et al. (2004) report

that within the family system, siblings are particularly helpless because they may be the least directly involved with the treatment process of the ill child, but their lives may still be affected by changes in daily routines and such. The lack of control felt by siblings can negatively affect their emotional well-being and forces them to rely on various coping strategies in order to adapt, which may include emotion-focused or cognitive coping strategies (Houtzager et al., 2004). For example, the sibling can try to trust that the medical specialists are competent and to believe in the medical treatment, remain hopeful that the situation will improve, or comprehend the situation in order to gain a sense of control (Houtzager et al., 2004). These coping strategies impact adjustment, thereby impacting quality of life. Houtzager et al. (2004) conducted a study to determine the extent of coping strategies and family cohesion on the psychological well-being of siblings of children with cancer. Eighty-three siblings between the ages of 7 and 18 participated. One or two siblings from each family were included. Half of the siblings were older than the ill child. The researchers measured the psychological well-being of siblings using self-reports of quality of life, anxiety, emotional-behavioral problems, and illness-specific emotional reactions. Parent proxy reports of psychological well-being were assessed, as well (Houtzager et al., 2004). The findings indicated that female siblings experienced more anxiety than male siblings and older siblings experienced more anxiety than younger siblings. Siblings of children with cancer also reported behavioral-emotional problems, as measured by the Dutch Child Behavior Checklist, that were associated with the fatality of the child's illness (Houtzager et al., 2004). Parents experiencing more stress than others due to their child's illness also rated their well child as having more psychological problems as measured by the Child Behavior Checklist

(Houtzager et al., 2004). The researchers also found that older siblings reported a lower quality of life than younger siblings of ill children. Self-report of behavioral-emotional problems was associated with the fatality of the affected sibling's illness and with level of family adaptation to the illness (Houtzager et al., 2004). Siblings of children with LQTS may adapt better or worse than siblings of children with other chronic illnesses and disabilities. Factors such as overall family adjustment, knowledge of the illness, and age may determine the impact on quality of life of siblings of children with LQTS, as shown in other studies of siblings of children with chronic illness.

Quality of life.

According to Bonomi, Patrick, Bushnell, and Martin (2000), the World Health Organization states, "quality of life can be viewed as a subjective, multidimensional concept, which places emphasis on the self-perception of an individual's current state" (p. 1). Quality of life includes health status, psychological well-being, and social functioning (Bonomi et al., 2000). In addition, quality of life incorporates level of independence, personal beliefs, and relationships to features of the environment (Bonomi et al., 2000). Quality of life can also be defined by the notion of a good life, as explained by the integrative quality of life theory.

As stated by Ventegodt, Merrick, and Jorgen Anderson (2003), quality of life is linked to one's culture. The Western cultures view a good life as one that includes happiness, fulfillment of needs, meaning in life, well-being, satisfaction with life, biological order (physical health), realization of one's potential, and objective factors such as income, marital status, and contact with others (Ventegodt et al., 2003).

According to the integrative quality of life theory, quality of life can be separated into

subjective, existential, and objective. The subjective quality of life is each individual's evaluation of a good life or a life of high quality. An individual evaluates how he or she views the world and his or her feelings about various things. The existential quality of life is the evaluation of a good life on a deeper level; an individual evaluates whether he or she is living in accordance with spiritual or religious values. The objective quality of life is the person's life as perceived by others in the culture in which he or she lives (Ventegodt et al., 2003). A sibling's quality of life may be affected by the quality of life of the family as a whole. As indicated by Poston and coworkers (2003), the concept of family quality of life encompasses the degree to which the family members' needs are met, the extent to which they enjoy spending time together, and the degree of engagement in activities they enjoy doing together. In the case of a sibling of a child with LQTS, quality of life could be impacted by the attention received from his or her parents, the relationship with the ill child, and the way in which his or her parents perceive the sibling in relation to the child with the syndrome.

According to Wrosch and Scheier (2003), a good quality of life indicates successful growth as a person. A good quality of life also indicates that the hopes of an individual have been matched by his or her experiences. A poor quality of life, however, occurs when an individual's hopes do not match his or her experiences (Skevington, O'Connell, & the WHOQOL Group, 2004). If a sibling of a child with LQTS has been negatively affected by the illness in terms of limited parental attention, an unwelcome increase in responsibilities, or feelings of isolation and worry, quality of life may decrease. Effective coping skills that help siblings adapt better to the changed circumstances in the family may help prevent psychosocial problems, thereby limiting the

negative impact on quality of life (Houtzager et al., 2005). Coping strategies can either be problem focused or emotion focused, depending on the situation (Houtzager et al., 2005). According to Lazarus and Folkman (1984), families and siblings are least likely to use problem focused coping in a situation that is out of their control, such as the diagnosis of chronic illness. The ill child and members of his or her family are most likely to use emotion focused coping, which focuses on resisting any threats to the family dynamic and unpleasant feelings that may arise (Lazarus & Folkman, 1984). Negative feelings associated with the diagnosis of chronic illness can be reduced by trusting the medical staff, maintaining a positive view of the process, and understanding the illness (Houtzager et al., 2005).

Quality of life, as defined by the World Health Organization, may be positively or negatively influenced by the birth of a child with a chronic illness. If siblings of children with chronic illness or disabilities are vulnerable to poor psychosocial outcomes, including an increase in emotional and behavioral issues (Breslau et al., 1981; Lavigne & Ryan, 1979), then it is important to address these outcomes in order to increase the chances for a good quality of life (Bellin & Kovacs, 2006). Although research has shown both positive and negative effects of chronic illness on siblings, no available research has shown the effects of long QT syndrome on the quality of life of siblings.

Developmental stage.

As indicated by Piaget, as children reach school age, they become better able to recognize other people's perspectives, as there is a loss of egocentric thinking (Sigelman & Rider, 2009). Therefore, starting at the age of 7 or 8 years, children can sense distress in a parent and model that method of adjusting to the diagnosis of a chronic illness. In

addition, concrete and formal operational stages are characterized by feelings resulting from interactions with others (Carroll & Steward, 1984). In a study conducted by Carroll and Steward (1984), children in both the preoperational stage and the concrete operational stage of development were provided with various feelings tasks. Children in the preoperational stage used cues from situations to respond to questions about their feelings, while children in the concrete operational stage of development referred to internal experiences apart from the situation to describe their feelings.

In summary, the diagnosis of chronic illness or disability in a family can have a positive or negative impact on each member, including siblings. Positive impacts on siblings include an increased sense of empathy for others and the ability to problem solve in stressful situations. Negative impacts include increased irritability and a feeling that the ill child is receiving more attention than the well child. Poor psychosocial outcomes for siblings of children with chronic illness may lead to a decrease in quality of life, as indicated by how they view life and how others view them. Long QT syndrome is a chronic illness that can lead to syncope, cardiac arrest, and death. Research on the impact of long QT syndrome on the family, particularly siblings is limited.

Research question

This research aimed to identify the ways in which LQTS may affect a sibling without LQTS. The research question is: How does long QT syndrome affect the siblings of individuals with LQTS?

Chapter 2: Method

A qualitative research design was utilized to determine if and how LQTS in children impacts quality of life of unaffected siblings. Siblings of children with LQTS participated in a semistructured interview or completed an online survey with open-ended questions.

Design and Design Justification.

Although there is a great deal of research examining the social, physical, and psychological adjustment of both the families and siblings of children with disabilities and chronic medical illness, no available research has focused solely on the functioning of siblings of children with long QT syndrome. A qualitative design was chosen to understand how LQTS impacts the unaffected siblings of children with long QT syndrome and reveal their experiences without restrictions and assumptions (Kazdin, 2003). Participants openly discussed their experiences of having a sibling with LQTS. Multiple methods of recruitment and data collection enhanced the exploration of the topic. As indicated by Baxter and Jack (2008), multiple data sources allow the researcher to study the phenomenon through a variety of lenses, ensuring that many facets of the occurrence are revealed and understood. Overall, qualitative researchers focus on *how* things occur rather than on the occurrence (Tetnowski & Damico, 2001). The focus was on how LQTS impacts an unaffected sibling in terms of social, physical, and psychological adjustment.

Data Analysis.

The current study utilized the qualitative method of research, which led to the emergence of themes that were used to explain the phenomenon of interest (Kazdin, 2003). The study utilized both face-to-face interviews and an open-ended, Internet-based survey with siblings of children with LQTS in an effort to explore and describe the unaffected siblings' experiences of living with a child with the disorder. Grounded theory was used to develop the aforementioned themes. Grounded theory is most often used to generate hypotheses, rather than to test established hypotheses (Auerbach & Silverstein, 2003). Grounded theory allowed the researcher to use her own concepts derived from the interviews and surveys in an effort to describe the experience of the unaffected child (Glaser, 2002). After themes emerged, ideas about an applicable theory to the population being studied became more apparent.

The grounded theory method utilizes a data analysis process called *theoretical coding* (Auerbach & Silverstein, 2003). Theoretical coding is used to generate hypotheses based on the information obtained from the participants throughout the interview (Auerbach & Silverstein, 2003). Coding allowed the researcher to review transcribed face-to-face interviews and survey responses, identify text relevant to the research question, and utilize repeating ideas to create themes. The themes were then used to generate more abstract ideas known as theoretical constructs (Auerbach & Silverstein, 2003). Coders included the researcher and two other graduate students who have knowledge and experience with long QT syndrome. All coders were supervised by a licensed psychologist with experience in qualitative methodology. During initial review of the recordings and survey responses, each coder engaged in process note taking

or memoing, wherein they gained insight into emerging ideas and generated initial hypotheses about the participants' experiences of living with a sibling with LQTS. Memoing was used to aid the researcher and coders in making the connection between the raw data and the phenomenon being studied (Birks, 2008). Initial review of the recordings and survey responses allowed the coders to understand the world from the perspective of those being interviewed and surveyed (Corbin & Strauss, 2008).

Coding strategies used included open coding, axial coding, and selective coding. During the initial review of the transcribed interviews and survey responses, the coders utilized open coding to assign codes to words or phrases that appeared relevant to the research question. Axial coding was then used to compare open codes with each other in order to create categories. Lastly, selective coding was utilized to group frequently occurring axial codes into core categories, or theoretical constructs (Edwards, Huebner, Connell, & Patrick, 2002).

Sampling, consensual validation, and saturation were considered when coding. In grounded theory method, the sample size cannot be determined in advance (Auerbach & Silverstein, 2003), but it was thought that 10 participants would be sufficient. Therefore, the researcher attempted to continue interviewing participants and posting the survey until the responses ceased to produce new information that added new concepts to the theory (Auerbach & Silverstein, 2003). Hearing the same information repeatedly indicates that the sample size is sufficient and saturation has been reached. For consensual validation, it was important that the coders agreed on particular meanings attributed to emerging themes and theoretical constructs. In order to ensure consensual

validation, all coders reviewed all transcripts and survey responses and discussed codes assigned individually until a consensus was reached.

Participants.

Setting.

The study data was collected at the homes of two participants and at the Philadelphia College of Osteopathic Medicine (PCOM) for a third participant. The interviews were scheduled at times convenient for both the researcher and the study participants. The anonymous, Internet-based survey was posted to social networking sites and completed anonymously by adult participants at their leisure.

Recruitment.

Initially, the researcher intended to recruit 10 children and adolescents ranging in age from 8 to 18 years. Participants were recruited from the Cardiac Center at the Children's Hospital of Philadelphia. A letter describing the study was mailed to 45 prospective participants, along with a return postcard and e-mail address used by the researcher to confirm interest in participation. Letters and postcards were sent to potential participants twice, with approximately 6 months between mailings. Information about the study was also sent to local area hospitals and doctors who treated potential participants. Potential subjects who live in the mid-Atlantic region were also recruited online from LQTS informational websites and other Internet sites, such as Craigslist, Facebook, and Twitter. Participants were also recruited by word of mouth and snowball sampling. Prospective participants informed the investigator of their interest by returning the postcard or via e-mail. Upon receiving the postcard or e-mail, the investigator contacted the parent of the potential participant to discuss the study, answer any

questions, and schedule the interview. Three participants, all children, were recruited from the first mailing of letters and postcards. These three participants were each given two movie tickets to thank them for participating. Due to limited participation, recruitment was expanded to potential subjects over the age of 18 years via an Internet-based survey posted on LQTS informational websites and other Internet sites, such as Craigslist, Facebook, and Twitter. An e-mail was also sent to past participants of other PCOM LQTS studies. Four adults were recruited and anonymously completed the Internet-based survey. If the respondent was interested in receiving two movie tickets for participating, he was asked to e-mail his contact information to the researcher.

Inclusion Criteria.

Participants in the study were unaffected biological siblings of children diagnosed with long QT syndrome for at least 1 year, regardless of severity. The researcher originally intended to include only siblings from 8 to 18 years of age. The face-to-face interviews were conducted with child participants aged 9, 12, and 13 years. Due to a poor response to mailed letters, an Internet-based survey was used to collect additional data from participants 18 years of age or older. The unaffected siblings resided in the same household as the children with long QT syndrome or previously resided in the home with the affected child prior to the age of 16 years. The unaffected siblings had to know that his or her sibling has long QT syndrome. The unaffected siblings participated in the interview without parents present, but with parental consent and assent. The participants were English speaking and could be from either a two-parent or single-parent family. Initially, the study required that participants reside within the tristate area to facilitate

face-to-face interviews. In an attempt to expand recruitment, the Internet-based survey did not require United States residency.

Exclusion Criteria.

Exclusion criteria included a LQTS diagnosis of less than 1 year. Families with siblings not residing in the same home presently or prior to the age of 16 years were excluded from the study. Siblings of children with LQTS with a new ICD implant or sudden cardiac arrest within the last 6 months were also excluded. In addition, siblings with a medical illness or physical disability were excluded from the study. Siblings with a mental health diagnosis that precluded them from being involved, at the discretion of their parents, were also excluded. Inclusion and exclusion criteria were verified by screening by the researcher before the interview or a list of questions prior to beginning the Internet-based survey.

Demographic characteristics.

A personal information questionnaire (Appendix A) was completed by one parent of each child subject who participated in the face-to-face interview. A total of three interviews were conducted. An abbreviated version of the personal information questionnaire was also completed for the Internet-based survey (Appendix B) by four individuals over the age of 18 years. The table below provides demographic information.

Table.

Demographic Characteristics

Identifier	Age	Gender	Birth Order	Parent(s) With LQTS
Child000	13	Male	Twin	No
Child001	9	Male	Youngest	Mother
Child002	12	Male	Oldest	No
Adult1	>18	Unknown	Oldest	Father
Adult2	>18	Female	Youngest	Mother
Adult3	>18	Unknown	Youngest	Mother
Adult4	>18	Female	Oldest	Mother

Measures.

Face-to-face interviews were conducted with unaffected siblings under the age of 18 years of children with long QT syndrome. While the interviews were being conducted, a personal information questionnaire regarding the participant was completed by a parent. The information requested included number of friends, engagement in social activities, performance in school, and the quality of the relationship with the affected sibling. Interview questions (Appendix C) were developmentally appropriate and included knowledge of LQTS and observations made by the siblings regarding their parents and the affected child. An Internet-based survey using the same questions was posted online (Appendix D). These surveys were completed by unaffected siblings over the age of 18 years of children with LQTS. A briefer version of the personal information

questionnaire (Appendix A) was also included in the Internet-based survey and included questions pertaining to social activities and performance in school.

Protection of Human Subjects.

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

Procedure.

Parents of potential participants under the age of 18 years were screened via telephone to determine if the children were eligible. Following selection of participants, the researcher arranged to meet participants in their homes or a mutually agreed upon location during convenient times for the parents. The researcher and the participants met privately in order to maintain confidentiality and promote honesty in answering the interview questions. Time was spent at the beginning of the interview establishing rapport with each participant. Each interview lasted approximately 1 hour. Each participant was asked both closed- and open-ended questions about perception of the impact of LQTS on self, parents, and the affected child. The researcher asked for clarification where needed. All interviews were audio recorded, with the permission of both the parents and the participant.

Due to a low response rate to the letters, an Internet-based survey featuring both open- and closed-ended questions was created to obtain more data. Participants were required to be over the age of 18 years and participation was anonymous; obtaining consent was therefore not required. Prior to beginning the survey, each participant completed screening questions. If inclusion criteria were not met, the potential participant was disqualified.

Chapter 3: Results

Analysis of the data collected resulted in two emergent themes: relationships and strategies of coping. Relationships between the participants and their parents, affected sibling, and/or friends/support systems were discussed, as were strategies of coping. Relationships and coping strategies may be explained by developmental stage of the participants, as well as awareness of the syndrome, which are included as considerations for the findings. These two themes provided the researcher with insight into the impact of having a sibling with LQTS.

Interviews.

Three face-to-face, semistructured interviews were conducted. All participants were English-speaking, Caucasian males and ranged in age from 9 to 13 years. Participants did not have LQTS. All three participants had one sibling with LQTS; however, their birth positions varied: one participant was a twin, one participant was younger, and one participant was older than the affected sibling.

Prior to asking the interview questions, the researcher established rapport with the children, inquiring about their likes/dislikes regarding activities, food, school subjects, and television shows. All three participants enjoyed both watching and playing sports. Likes/dislikes in regards to food, school subjects, and television shows varied. However, all three subjects identified writing as their least favorite subject in school.

Internet-based surveys.

Of the 41 responses received for the Internet-based survey, only four participants met the criteria and completed the survey in its entirety. A fifth person completed the

survey, but reported having LQTS, which was an exclusion criterion. Therefore, this data was not used. All participants were 18 years of age or older and English speaking. All respondents reported they did not have LQTS and reported being the biological sibling of a child with LQTS. Some of the survey questions asked for responses about the participants' childhoods.

Themes.

Key themes were identified following coding and analysis of both interview and survey data from the seven participants. The themes were discussed and agreed upon by the researcher and two coders following individual and group review of the data collected.

Relationships.

Parent-child relationship.

Among the seven participants, five believed that the relationship with either or both parents changed after their siblings were diagnosed. For the most part, the unaffected siblings felt that the parents became more protective of the children with LQTS, possibly due to physical restrictions. For example, some of the affected siblings had to drink a certain amount of water per day and avoid certain types of food. Some of the participants expressed their understanding of why their parents may be more protective and concerned about the affected child. Other participants felt that their parents favored the affected sibling more, in that they were catered to or provided with more slack than the unaffected sibling. One participant attempted to explore other reasons that the affected sibling may be provided more slack, such as having the role of little sister. In retrospect, some of the adult participants reported that their parents either

expected the unaffected sibling to take on more responsibility or became so distracted with the affected child's illness that the unaffected sibling was no longer a priority.

Although the relationships between the unaffected siblings and the parents were discussed briefly, responses to other answers helped to provide more insight into those relationships. For example, all of the participants reported participating in extracurricular activities, particularly sports. Active participation in sports indicates that the unaffected siblings were not restricted from social activities, although the affected siblings may have been unable to participate due to LQTS. This indicates that parents of the unaffected siblings respected their individuality and personal interests; the affected siblings were not limited in engaging in activities they enjoyed simply because the affected siblings may have been unable to do so.

Unaffected sibling-sibling relationship.

Three of the adult participants noted a change in the relationship with the affected siblings following diagnosis. Two of the adult participants reported becoming more protective of their siblings following the diagnosis, reminding the sibling to take medication and just being more "motherly" towards the affected sibling. The third adult participant reported feeling excluded because more than one sibling was diagnosed with LQTS. One of the adults and the three children did not report any differences in the relationship since diagnosis.

A majority of the participants appeared to be quite protective of the affected sibling. Although many of the participants reported attending at least one medical appointment with their sibling, one participant reported wanting to attend so as to be directly involved with treatment. Another participant reported feeling like it was her job

to make her sibling smile, as the affected sibling was very unhappy with the various restrictions placed on her following diagnosis. Two of the child participants discussed social situations related to their siblings' LQTS. For example, one participant reported overhearing other children talk about his brother behind his back, while another participant discussed how his sibling feels weird and different because she has an aide at school. A majority of the participants appeared to feel sympathy for their siblings and a desire to protect them from the outside world.

Only two participants reported not getting along with the affected siblings. One felt left out, as most of her family was affected by LQTS; therefore, the participant felt isolated from the affected siblings "sharing something so big." Another participant thought the affected sibling was more successful, which she attributed to making poor decisions as a teenager after feeling isolated from the family following the LQTS diagnosis. A majority of the participants discussed a typical sibling relationship, with one participant stating that the worst thing about his sibling having LQTS is that she has a pacemaker and he is not able to roughhouse with her as much as he would like.

Emotional considerations.

A majority of the participants expressed concern and worry about the affected siblings. The unaffected siblings reported reminding the affected sibling to take medication and avoid certain activities. For example, in discussing physical limitations of LQTS, one participant stated that he worries about his sibling when he "pushes himself...over his limits." He reported often reminding his sibling to take rest breaks for fear of what could possibly happen. Another participant reported feeling badly for his sibling because she is missing out on doing some things.

Sibling-friends/support system.

Although briefly discussed, all of the participants had friends outside of the family unit. In addition to having friends, all of the participants reported being involved in extracurricular activities outside of school (i.e., sports). For the child participants, the researcher asked the parents to complete a personal information questionnaire for the unaffected participant (Appendix A). The parents indicated that the participants had at least 2 or 3 close friends. Each participant had also spent nights away from home at a friend's house. Being involved in extracurricular activities and sleeping away from home suggests that the unaffected siblings are not socially restricted, even if their siblings are unable to engage in these types of activities. For the most part, many of the participants' friends know about the affected siblings' illness, but do not usually discuss it or ask questions unless they witness an event related to LQTS. For example, one adult participant stated that her friends know about the LQTS and asked about it when the sibling passed out at a school related sports event because it was a big deal among their peers.

All participants were asked with whom they can share their feelings. A majority of the participants were able to identify at least one person they feel comfortable sharing their feelings with, either in person or by phone, e-mail, or text message. This indicates that the siblings have an outlet to vent if they were to feel negatively about their siblings' illness, family's reaction to the illness, or anything else. Most of the child participants reported having friends/family members in whom they feel comfortable confiding. Four participants identified their mothers as the person they would speak to if they needed to talk about how they were feeling.

Coping strategies.

A diagnosis of LQTS can be stressful for the affected child, as well as for the family. As indicated by Lazarus and Folkman (1984), coping refers to an individual's actions that are focused on handling stress. Coping is a process with no prescribed good or bad actions, but ultimately ends in adaptation rather than consequences related to poor adjustment (Lazarus & Folkman, 1984). An individual can cope behaviorally, taking actions to solve the problem, or focus coping on emotional regulation or control (Houtzager et al., 2004). Problem-focused coping involves taking action to modify the problem and includes generating options to solve the problem, weighing the pros and cons of each option, and implementing steps to approach and solve the problem. Emotion-focused coping usually aims to manage the negative feelings associated with the problem (Lazarus & Folkman, 1984).

Emotion-focused coping.

According to Houtzager et al. (2004), there is little the family can do to change or exert any direct control over the situation when a family member is diagnosed with a chronic or life-threatening illness. In this case, some unaffected siblings may engage in more cognitive or emotion-focused coping strategies than problem-focused strategies as a result of feeling helpless because they are typically the family members least involved in the treatment process (Houtzager et al., 2004). Emotion-focused coping strategies can range from denial and avoidance to venting of emotions, seeking social support, and positive interpretation of the situation. The effectiveness of emotion-focused coping tends to depend on the type of strategies used (Carver, Scheier, & Weintraub, 1989).

In the current study, a majority of the unaffected siblings appeared to cope by placing their trust in the affected siblings' doctors and the recommended treatments. For example, one participant noted that the affected sibling took "gross" smelling medication, but appeared confident that his sibling was doing well and did not think much else about his sibling having LQTS. Other participants reported taking on more responsibility, reminding the affected sibling to take medication, possibly indicating trust in and dependence on pharmacological treatment. In fact, one participant reported that the affected sibling had not had any of these episodes since starting the medication 14 years ago.

Some of the participants, mostly the adults, also appeared to be less anxious about the illness when they had more information. These participants were very matter of fact in their thoughts, based on their knowledge of the illness, as well as the prescribed treatments. For example, when one participant was asked how she feels about the affected sibling wearing a heart monitor, she stated, "I know that it is part of the treatment, so it doesn't really make me feel strongly." Another participant reported feeling sad about her sibling having LQTS; this sibling was the only one who reported attending the affected sibling's medical appointments because she wanted to be involved in the treatment. It is possible that this participant is seeking more knowledge about LQTS in order to deal with the negative emotion of sadness.

Summary of results.

In coding the data, two themes emerged regarding the effect that the diagnosis of LQTS has on the unaffected sibling: relationships and coping strategies. The relationships with one's parents, affected sibling, and friends can be affected by the

diagnosis of chronic illness in the family. The current study found that, for a majority of the participants, relationships with the parents changed after diagnosis. Most of the participants reported that the parents became more protective of the affected sibling, at times catering to the affected sibling or providing the affected sibling with more slack. Some of the adult participants also noted an increase in responsibility following diagnosis, although they did not specify in what ways they were expected to be more responsible. In regards to relationships with affected siblings, a majority of the participants did not feel that their relationships with the affected siblings changed. However, some of the participants reported feeling more protective of the affected sibling. All of the participants expressed some degree of concern and worry about the affected sibling. All of the participants felt that they had supportive people in their lives.

In regards to coping, it appeared that a majority of the participants had a sense of vicarious control, placing their trust in the medical team and treatments prescribed to the affected siblings. In addition, participants also appeared to have a sense of interpretive control, as they had knowledge of the illness, as well as the treatments and restrictions related to the syndrome.

Chapter 4: Discussion

In the current study, the experiences of having a sibling with long QT syndrome were examined. Previous research has explored how parents cope with their children's diagnosis of LQTS, and how the affected child copes with and manages living with the illness (Burns-Pentecost, 2013; Gonzales, 2009; McElwaine, 2015). In the current study, participants discussed and expressed their experiences of having a sibling with LQTS. The two themes that emerged were relationships and strategies of coping.

Theoretical analysis of findings.

The diagnosis of a chronic illness can be quite disruptive, as it can pose a persistent threat to the structure and functioning of the family (Cohen, 1999). Chronic illness can disrupt individual and family emotional well-being (Houtzager et al., 2004). How the family as a whole adjusts to and copes with the illness can impact how each individual member adjusts to and copes with the illness. Research has shown that childhood chronic illness has an impact on well siblings; parents may have less time to spend with and nurture the well siblings in their development and/or parental distress may also affect the well siblings (Cohen, 1999). It is for this reason that a family systems approach to the investigation of the impact of LQTS on the unaffected sibling is warranted.

The family as a system.

As discussed previously, the family unit can be viewed as a system in which there is a whole with interdependent parts. Each element of the system, or member of the family, relies on the others in order to function. Also, as in certain systems, there is a circular rather than linear pattern of involvement in the family unit. For example, it is

difficult to say whether overprotective parents create anxiety in their children related to the illness. It could be that the parents and children have created a pattern where the fears of the children trigger the parents, which makes the anxiety of the children worse. For example, all of the participants in the current study expressed concern and worry about the affected sibling; however, 3 of 7 participants actually witnessed an event related to LQTS, possibly indicating that the concern and worry may stem from parental worry about the diagnosis and potential symptoms. The worry may also be a result of the siblings' own knowledge of the illness.

Systems also have features that work to maintain homeostasis. When behaviors depart from what is expected within the family, the family can work to correct those behaviors and reclaim homeostasis. Families do not plan on the diagnosis and management of a chronic illness; when a chronic illness occurs, the family must adapt its daily functioning. As indicated by Cohen (1999), *balanced coping* refers to a protective strategy families may use to meet the needs of the illness while maintaining the family life cycle. As indicated in the current study, the unaffected siblings were still able to engage in social and recreational activities, although the affected siblings may have been restricted. Perceived catering to the affected siblings may have been the parents' attempts at trying to balance the needs and wants of all siblings. Finally, in systems, evolution and change are expected (Minuchin, 1985). Therefore, when one family member's functioning changes, another family member automatically compensates for that change (Bowen, 1974). For example, when one member of the family becomes ill, another member may compensate for the change by overfunctioning within the family. If the sick family member becomes chronically ill, a long-term imbalance in the family may

result from the overfunctioning of other family members. Some of the participants, specifically those over 18 years of age, indicated an increase in responsibility following a diagnosis of LQTS, although they did not describe the capacities in which their responsibilities increased.

The family system is composed of subsystems, including the spouse subsystem, the parent subsystem, the parent-child subsystem, and the sibling subsystem (Minuchin, 1985). Each subsystem is separated by boundaries, and interactions across those boundaries are determined by rules and patterns (Minuchin, 1985). If homeostasis is disrupted when a child is diagnosed with a chronic illness, the rules and boundaries regarding interactions between the subsystems must change. A majority of the participants noted a change in the relationships with their parents following diagnosis. Some participants noted a favoritism towards the affected child, which was perceived as necessary by one participant and as being more overly cautious by another. This may indicate that some of the rules and boundaries of that particular subsystem have been affected and not yet redefined to allow better adjustment to the changes that the chronic illness has caused in the relationship. A majority of the participants reported no changes in the relationships with their affected siblings after the diagnosis. Some participants did note feeling more motherly or protective of the affected siblings, which may or may not be viewed as a positive change in rules and boundaries within the sibling subsystem relationship.

Biopsychosocial systems approach.

To make the family systems approach more applicable to families facing chronic and life-threatening illness, researchers combined it with the biopsychosocial theory to

create the *biopsychosocial systems approach* (McDaniel & LeRoux, 2007). This approach aims to address the biological, psychological, and social/interpersonal issues in a family that may be caused by or affect chronic illness. The focus is on medical issues, as well as the body, mind, and family; this approach recognizes the biological features of a psychosocial problem and the psychosocial features of a biological issue (Doherty, McDaniel, & Hepworth, 2014; McDaniel & Hepworth, 2004). As indicated by McDaniel, Campbell, and Seaburn (1995), equal attention should be given to the affected person and family as members of the treatment team. Affected siblings and their families are considered the experts on their experiences and often the creators of their own solutions when problems arise due to the illness.

The biopsychosocial systems approach is utilized in medical family therapy for families living with chronic illness. Medical family therapy focuses on the strengths of the family, as well as the role of the illness in the lives of the patient and family (McDaniel & Hepworth, 2004). Utilizing this approach, the two goals of medical family therapy are increasing agency and communion. Most often, these terms refer to the affected patient. However, both terms can also apply to the unaffected siblings adjusting to and living with the diagnosis of LQTS in a sibling. First described by Bakan in 1966 and revised over the years, *agency* and *communion* refer to autonomy in relation to a specific context. Agency refers to a sense that one can make choices for himself or herself, while communion refers to connectedness to others (Doherty, McDaniel, & Hepworth, 2014). In other words, communion can refer to the unaffected siblings' abilities to maintain relationships following diagnosis and current management of LQTS,

while agency can refer to how the unaffected siblings cope with having a sibling with the illness.

Relationships/Communion.

As reported by Fife, Norton, and Groom (1987), there is an association between the quality of the parent-child relationship before a diagnosis and the occurrence of psychosocial problems following a diagnosis of a chronic or life-threatening illness. A majority of the participants noted changes in the relationships with their parents; however, the differences had more to do with being more protective and catering to the affected sibling a bit more due to physical limitations and treatments than with being completely disregarded as an individual. Other changes noted included being expected to take on more responsibility following the diagnosis; according to the family systems approach, this may be a necessary role change to maintain family functioning. Although some participants may have had additional responsibilities in the home, all participants were allowed to engage in social activities, which were most often sports related. Many of the participants also appeared to be knowledgeable about LQTS, indicating that their parents shared information with them appropriately. For a majority of participants, it appeared that the parent-child relationship established prior to the diagnosis appeared to remain intact.

A majority of the participants did not report any differences in the sibling subsystem relationship following diagnosis of LQTS. Most of the siblings appeared to get along, and if they did not, it appears as though that was the nature of the relationship prior to diagnosis. One participant reported having two siblings diagnosed with LQTS, leaving her as the only sibling without the illness. This sibling subsystem relationship

was negatively affected, as the unaffected sibling felt isolated. In regards to relationships with friends and other social supports, most participants were able to identify people both within and outside of the family whom they could talk to if they needed support. Overall, connectedness appeared to be maintained throughout all subsystems, as well as in the community.

Strategies of coping/agency.

As indicated by Houtzager et al. (2004), when a family member is diagnosed with a chronic or life-threatening illness, there is little the family can do to change or exert any direct control over the situation. In this case, some unaffected siblings may feel helpless because they are typically the least involved family members in the treatment process; therefore, unaffected siblings may engage in more cognitive or emotion focused coping strategies than problem-focused strategies (Houtzager et al., 2004).

Rothbaum's two-process model of perceived control.

As indicated by Rothbaum et al. (1982), people value the perception of control. Both helplessness and locus of control theorists interpret behaviors such as passivity and withdrawal as giving up control; in other words, these behaviors are considered maladaptive. However, these behaviors can also be perceived as aligning with environmental forces when a person is unable to bring the environment into alignment with what he or she wants and needs, which is akin to active problem solving (Rothbaum et al., 1982). Therefore, the use of emotion-focused coping or secondary control may be used by some unaffected siblings who feel unable to exert any direct control over the illness and the ways in which it changes family functioning. In an attempt to adjust to resistance from a problem that cannot be solved, an individual may engage in four types

of control that feel safer: predictive control, illusory control, interpretive control, and vicarious control. Predictive control refers to efforts made to maintain a positive outlook. Vicarious control refers to the perception that others can control the situation. Interpretive control refers to efforts to understand the illness and derive meaning from it, which reduces feelings of ambiguity about the illness. Lastly, illusory control refers to wishful thinking and possibly belief in the control of a greater power (Rothbaum et al., 1982). The current study found that a majority of the participants utilized either or both secondary coping strategies of interpretive and vicarious control. A majority of participants appeared to put their trust in the affected siblings' physicians and in the treatments prescribed (vicarious control). Those with more knowledge about LQTS also appeared to be less anxious about the illness, although they continued to express worry and concern for the affected sibling (interpretive control). Because chronic illness is not something that can be changed, only managed, unaffected siblings of those with LQTS cope by engaging in some form of perceived control, whether it is trusting in the physicians or learning information about the illness. Unaffected siblings are choosing how to cope with the illness, which is in line with the concept of agency.

Resilience.

Resilience evolves with development; no one characteristic can be identified as a protective factor universally (Rutter, 1993). The only facet of resilience that can be agreed upon is that it emerges from the exchange of risk and protective factors. Risk factors create or maintain the problem, while protective factors eliminate or decrease any negative consequences associated with the problem (Bellin & Kovacs, 2006). The literature suggests that siblings of children with chronic illness are vulnerable to poor

psychosocial outcomes. Risk factors include parental distress, changes in family structure and roles, and decreased physical and emotional availability of caretakers (Cox et al., 2003). Other research has demonstrated positive effects, including increased empathy and an ability or willingness to assume more responsibility (Havermans et al., 2015). There has been little research on resiliency factors in this population. The research that has been conducted suggests that resiliency comes from individual protective factors, family protective factors, and environmental protective factors. Individual factors include the ability to communicate, being sociable, and having a high self-esteem (Masten & Coatsworth, 1998). A majority of the participants in the current study appeared to have a strong support system, both within and outside of the family unit. Those interviewed were also able to communicate fairly well and appeared to pride themselves on their performances in sports and/or academics, indicating high self-esteem. Family protective factors include a close relationship with at least one caretaker. A majority of the participants in the current study reported having a good relationship with at least one of their parents or another adult family member. Lastly, environmental protective factors include being involved in prosocial activities. A majority of the participants were involved in at least one social or recreational activity outside of school and the family. The current study found that, overall, the participants appeared quite resilient and did not experience negative outcomes other than feeling concern and worry about their siblings, most likely due to the protective factors mentioned above.

Quality of life.

As previously discussed, sibling quality of life may be affected by the quality of life of the family as a whole. The concept of family quality of life encompasses the

extent to which family members enjoy spending time together, and the degree of engagement in activities they enjoy doing together, and the degree to which their needs are met (Poston et al., 2003). Quality of life of a sibling of a child with LQTS could be impacted by the way in which the siblings' parents perceive the sibling in relation to the affected child, the attention received from the siblings' parents, and the relationship with the ill child. In consideration of the information provided, quality of life appears to be minimally affected in siblings of children with LQTS. Although some of their relationships were impacted, they overall appear to be able to engage in and enjoy most of what they need and desire to do.

A majority of the participants maintained positive relationships with their parents, the affected sibling, and had friends/support systems both within and outside of the home. Some participants did report an increase in responsibility or some favoritism for the affected sibling, but this did not appear to cause significant negative effects. All participants were involved in enjoyable activities and performed well in school. For the most part, the participants also reported a general positive mood state. A majority of the participants did not give the impressions that their lives differed greatly from siblings in other families not affected by LQTS.

Developmental stage as a consideration.

Child participants versus adult participants.

Among the seven participants in the current study, there were three children ranging in age from 9 to 13 years. The remainder of the participants were 18 years of age or older. The child participants provided responses that were focused on the present,

whereas the adult participants provided responses that were focused on both the past and present.

Erikson and psychosocial development.

According to Erikson, the child participants in the current study are somewhere between the stages of industry vs. inferiority and identity vs. role confusion (Sigelman & Rider, 2009). At this stage in their development, they are attempting to master social and academic skills in order to keep up with their peers in an effort to avoid feeling inferior (industry vs inferiority). They are also working towards establishing an identity, socially and vocationally, in an attempt to successfully enter their adult lives in the near future (identity vs. role confusion) (Sigelman & Rider, 2009). The child participants often compared themselves to the affected siblings in relation to sports and/or academics. One of the participants expressed frustration with his sibling who he felt was holding him back during track meets due to the physical limitations of LQTS. Another participant reported performing better in academics, although he and the affected sibling are rewarded equally. This stage of development may explain why some of the participants felt that their parents tended to cater to or favor the affected sibling. All of the participants discussed their involvement in social activities, indicating that peer relations were one of the most important aspects of life currently. Some of the participants also discussed future plans, including performing well athletically, but remained focused on the present for the most part.

Four of the participants were 18 years of age or older. Thus, there may be a large variation in stages of psychosocial development. The stages can range from identity vs. role confusion to integrity vs. despair. In other words, participants 18 years of age and

older may be struggling with their social and vocational identities (identity vs. role confusion) or intimacy and productivity issues and purpose in life (integrity vs. despair) (Sigelman & Rider, 2009). Stage of psychosocial development may explain how participants were able to look back on their experiences and have a different perspective on relationships than they may have had as children. One of the participants, currently a medical resident, discussed taking on the role of the responsible child, reminding the affected sibling to take his medication. Another participant reflected on the different path she took in relation to the affected sibling, stating that the sibling with LQTS has a successful life, leading the participant to regret some of the decisions she made as a direct result of feeling isolated in a family of LQTS patients. It is not only the psychosocial development of an individual that has an effect on relationships and coping styles, but the cognitive development, as well.

Piaget and cognitive development.

As indicated by Piaget, children lose egocentric thinking as they reach school age and are better able to recognize others' perspectives (Sigelman & Rider, 2009). Children in this age group can sense distress in their family members when adjusting to and navigating the diagnosis and treatment of a chronic illness. For example, the child participants in the current study explored how their siblings may feel about having LQTS. Two of the participants were able to express their sympathy for the affected siblings, as they understood the reasons for feeling different. In terms of problem solving, children in the concrete operational stage of cognitive development often take a trial and error approach, whereas individuals in the formal operations stage can think abstractly and hypothetically about problems (Sigelman & Rider, 2009). Piaget also indicated that

adolescents who reach the formal operations stage can also define justice abstractly. Two of the three child participants were most likely approaching the formal operations stage of cognitive development because they were able to discuss the idea of fairness more abstractly. For example, one of the participants reported that his parents catered more to the affected child, but for good reasons related to the LQTS diagnosis. He did not feel as though it was unnecessary to treat him differently from his sibling; therefore, the relationships with his sibling or parents did not appear to be negatively affected.

It is likely that a majority of the participants 18 years of age and older were in the formal operations stage of cognitive development. In addition to an ability to think abstractly and hypothetically about problems, those in the formal operative stage can also make predictions about the outcomes of their and others' actions. Siblings in this stage of development may be more protective of the affected siblings because they are more knowledgeable about the illness and know what can happen if it is not properly treated. For example, one participant expressed her concern about a sibling's future children and career choices. In terms of problem solving, other participants expressed an implicit obligation to take on more responsibility in order to make up for some perceived deficiencies in the affected siblings' ability to do so. In addition to affecting the type of relationships the participants have with their parents, siblings, and friends, developmental stage can also determine how the participants cope with and solve problems related to LQTS within the family.

A majority of the participants were at a cognitive stage at which they were able to understand others' perspectives and think about problems abstractly. These participants were also able to express sympathy for the affected siblings having to cope with and

manage LQTS. A majority of these participants were also able to understand why their parents became more protective or expected them to be more responsible following diagnosis. In regards to psychosocial development, some of these participants discussed their professions, performances in school, and/or sports and involvement with friends and other social activities (industry vs. inferiority and identify vs. role confusion), which implied attempts at differentiation from the affected siblings and the families. Through their responses, adult siblings demonstrated that the interdependence of the family had not restricted them from establishing an identity outside of the family system.

Awareness as a consideration of the findings.

Hendriks et al. (2005) found that parents of children with LQTS who were dissatisfied with information provided about the disorder experienced more distress than those satisfied with the information received. However, a qualitative study conducted by Hummelinck and Pollock (2006) explored information seeking by parents with chronically ill children and found that parents varied in the amount of information they desired. Some parents actively sought out information, but also resisted information as a coping strategy, fearing potential negative impacts. Sharing the information obtained with the affected child and unaffected siblings can be a difficult decision. Parents may have to decide what can be shared without inducing fear and worry in both affected and unaffected children. In another qualitative study conducted by Mangset and Hofman (2014), parents of children being tested for the LQTS gene felt that early and adjusted disclosure to the affected children would promote effective coping. These parents argued that all children have obstacles to overcome and this syndrome should not be looked at as something that will hold them back in life. Adults living with LQTS agreed that early

and gradual disclosure of information was an advantage in coping with the illness (Anderson et al., 2008).

The amount of information shared with the unaffected siblings may have an impact on the relationship with the parents and other members of the family. As indicated by Poznanski (1973), deficits in knowledge about an illness or disability may cause unaffected children to feel isolated and excluded from a significant family situation. All of the siblings had some knowledge about the illness and the treatments needed to control the symptoms (i.e., medication or an implantable cardioverter defibrillator). A majority of the participants also noted the physical restrictions associated with the disorder, including limited participation in sports and restrictions on amusement rides. Two of the three child participants had a vague knowledge of long QT syndrome. They both appeared to have a general idea of why the affected sibling takes medicine (“something to do with his heart”) and the siblings’ physical limitations. The third participant appeared to be more knowledgeable about LQTS, which may be because his parents were more open in discussing the illness. The adult participants appeared to be fairly knowledgeable about the illness.

Conclusion

The current study attempted to provide insight into the experiences of siblings of children with LQTS, as there is little prior research on siblings of children with LQTS. Through the current study, information was obtained about both the positive and negative aspects of being a sibling of a child diagnosed with LQTS, and the possible effects on sibling quality of life. The current study contributes to the literature by exploring the effects of the condition on siblings of the affected child. Overall, there appeared to be

minimal effect on quality of life of the unaffected siblings; however, each sibling described feeling concerned and worried at times, which warrants attention.

Understanding the effects on unaffected siblings may lead to an improvement in support and psychoeducation for those unaffected by LQTS. Specifically, families with LQTS need a biopsychosocial systems approach to their care, which would suggest an integrated interprofessional healthcare approach. Health psychologists working in medical settings should be available to families living with LQTS in an attempt to resolve any biological, psychological, or social/interpersonal issues, including academic, behavioral, or emotional problems that may arise while adjusting to the diagnosis and management of LQTS.

Limitations

Based on the nature of the research design, there are certain limitations of the study. As indicated by Kazdin (2003), qualitative research is not meant to describe all people; rather, the goal is to expand on the meaning and understanding of experiences in a specific context. However, it must be noted that the sample size was small, which limits the overall generalizability or transferability of the findings, although the themes found may have some universality among similar groups (Kazdin, 2003; Morrow, 2005). The sample size was homogeneous because all child participants were Caucasian males from upper middle class families, recruited through the same cardiology practice in Philadelphia, which further limits the generalizability of the findings. Another limitation is that the participants and their parents self-selected for participation in the study, possibly indicating that they were more well adjusted to the diagnosis, leading them to feel comfortable sharing their experiences.

A possible limitation specific to obtaining data from interviews may include self-report bias or social desirability that may have been present in face-to-face interview responses. As indicated by Kazdin (2003), subjects may alter the image of themselves in order to be represented in the best possible light. Participants in the interviews may have been more inclined to present themselves and their siblings and families in a favorable manner, which may have affected the data. The participants may have maximized the positive aspects of their experience and minimized any negative aspects. Experimenter bias may have also affected the data collected because the interviewer did not adhere strictly to the script. The researcher tended to follow the lead of the child participant and prompt where needed or request additional information from some participants, but not others, depending on their openness to answering certain questions.

There are also limitations specifically associated with collecting data via the Internet. Accessibility may have been a limitation because many eligible potential participants may not have had access to the Internet; therefore, they were unable to participate in the study (Nosek, Banaji, & Greenwald, 2002). There may have been additional sampling bias, as little was known about the characteristics of the respondents, other than some basic demographic information, and even this information may be questionable (Wright, 2006). In addition, there may have also been a response bias, as there were open-ended questions included in the survey; some participants appeared to understand the questions and others did not, which indicated that the questions may have not clearly explained what information was expected (Stanton, 1998). Also, there may have been motivational variability in participants, leading to additional response bias, as some respondents appeared to include a lot of information in their answers, while others

used yes or no responses only. The state of mind the respondents were in when answering the questions (e.g., intoxicated, moody, or tired) cannot be determined (Stanton, 1998).

Another limitation associated with qualitative research is related to threats to credibility, or internal consistency of the findings (Morrow, 2005). Credibility refers to the believability of the results; there should be agreement on interpretations made in an effort to enhance understanding of the phenomenon being studied (Kazdin, 2003). Due to the presence of experimenter bias in qualitative research, it is important to consult with other investigators in order to identify the extent to which the raw data reflects the key concepts and overall themes identified by the researcher (Kazdin, 2003). Therefore, multiple coders were used to increase interrater reliability or dependability of the findings (Morrow, 2005). However, it should be noted that each coder has his or her own perspectives and experiences, which may lead to coder bias in interpretation of the data.

Future research

Research on the experiences of siblings of children diagnosed with LQTS remains limited. Based on the current study, implications for new areas of research include determining additional protective factors that result in increased resilience in siblings of children with LQTS because they do not appear to be as negatively affected as siblings of children with other chronic conditions. For example, the cardiac specialist and parents may note that providing information to the unaffected siblings and/or allowing them to participate in office visits assist with coping and developing protective factors for resiliency. Research in this regard may identify ways to provide similar protective factors to increase resiliency for siblings of children with other illnesses and disabilities.

Due to the finding that unaffected siblings expressed concern and worry about their siblings, additional areas of research might include implementing ways in which siblings of children with LQTS can learn how to cope more successfully in an effort to decrease worry.

A follow-up study might incorporate a number of changes. First, the researcher should conduct in-person interviews with both child and adult participants in order to better understand their experiences. Also, after reviewing the transcripts and survey responses, missed opportunities to engage in more in-depth conversations and obtain more information were identified. In the future, researchers should ask follow-up questions in order to increase insight into and understanding of the sibling experience.

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**Appendix A: Personal Information Questionnaire (Interview) Personal Information
Questionnaire for Siblings**

Study ID# _____

Date: _____

Personal Information Sheet (Primary Caregiver)

Please **fill out** personal information questionnaire for your child:

Child's Age:

Child Gender:

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

Yes No

If yes, please describe...

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

Yes No

If yes, please describe...

SOCIAL ASPECTS

1.) 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? _____

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

None 1 2 or 3 4 or more

3.) 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

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

Yes If so, what? _____

No

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

Yes Please specify _____

No

TEACHERS AND SCHOOL SETTING

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

Home schooled Public School Private school

Other _____

7.) 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

8.) 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

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

Yes Please Specify_____

No

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

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

Yes

How many times?_____

Reason?_____

No

RELATIONSHIP WITH AFFECTED CHILD

12.) Please describe the relationship between the sibling and the affected child (i.e. play together or not, include each other in activities or not, speak kindly of each other or not, etc.).

INFORMATION REQUESTED ABOUT PRIMARY CAREGIVER AND/OR SPOUSE/PARTNER

12.) Age of Primary Caregiver _____

What is your relationship to the sibling? _____

Diagnosed with LQTS? Yes No

If so, symptomatic? Yes No

13.) Age of spouse/partner _____

Diagnosed with LQTS? Yes No

If so, symptomatic? Yes No

What is the spouse/partner's relationship to the sibling? _____

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

THANK YOU VERY MUCH FOR YOUR PARTICIPATION!

Appendix B: Personal Information Questionnaire (Internet-based survey)

What is your understanding of LQTS?

Besides your sibling (s), do you know anyone else with LQTS?

When you were of school age, were you involved in extra-curricular activities? If so, what?

When you were of school age, were you restricted from social activities you wanted to be involved in? Please specify.

When you were of school age did you sleep over at friends' houses or spend nights away from home with relatives?

How did you/do you perform in school? Any academic/behavioral problems? Please specify.

Have there been any significant changes in your life in the last year?

Appendix C: Interview Questions

1. Tell me a little about yourself (What are your likes/dislikes? What do you do for fun? What is your favorite subject in school? What is your favorite food/TV show/movie/song, etc.?)
2. (Establishing understanding of LQTS) We are all unique and great in our own ways. Each person needs a certain kind of care, care that is different. In what ways are you and your sibling different? In what ways are you similar? (PROMPT-Have you ever heard of LQTS (or what their parents may call it, which will be known prior to the interview)?/Why does your brother/sister see a heart doctor? What does that mean to you?
3. (How LQTS affects sibling) Can you think of anyway your brother/sister having LQTS (or what the parents refer to it as) affects you? Are there any good things about it? Are there any things that are less good about it? How does your sibling having LQTS advantage you/disadvantage you? Do you do more or less activities than your brother/sister? Does this ever come up between the two of you? What is your mood like normally? How do you feel about your sibling having LQTS? Do you think about it at all/even care? How do you feel when your brother/sister is sick or he/she has a heart doctor appointment/has to wear a heart monitor (Follow up question-Do you worry about your brother/sister? Why/why not?) Can you talk to anyone about your feelings? Who do you want to call, text, e-mail, or tell when something happens? Do you go to your sibling's heart doctor appointments? Why/why not? How is school? Do your friends know that your sibling has LQTS? Do they ask about it? (In parent questionnaire, will ask about grades/functioning in school)

4. (How LQTS affects sibling relationship) How do you get along with your brother/sister? Do you do things together?
5. (How LQTS affects relationship with parents) How do you get along with your mother/father? Has your relationship changed since your brother/sister started seeing the heart doctor? How do you think your parents treat your brother/sister? Are you treated the same or different? If different, provide examples.
6. Is there anything else I did not ask that you wanted to tell me/think is important for me to know?

Appendix D: Interview Questions (Survey)

What is your birth order? (Ex. Are you the first born? Second born?)

In what ways are you and your affected sibling(s) similar?

In what ways are you and your affected sibling(s) different?

Why does your affected brother(s)/sister(s) see a heart doctor?

What does it mean to you that your affected sibling(s) see(s) a heart doctor?

How did/does your sibling(s) having LQTS affect you?

Were/are there any good things about your sibling(s) having LQTS?

Were/are there any negative things about your sibling(s) having LQTS?

How did/does your sibling(s) having LQTS advantage you?

How did/does your sibling(s) having LQTS disadvantage you?

When you were of school age, did you participate in more or less activities than your sibling(s) with LQTS?

Did/does your sibling having LQTS ever come up between the two of you?

Describe your usual mood.

How did/do you feel about your sibling(s) having LQTS?

How did/do you feel when your affected sibling(s) has to wear a heart monitor?

How did/do you feel when your affected sibling(s) has a doctor's appointment?

How did/do you feel when your affected sibling(s) is sick?

Who do you talk to about your feelings, if anyone? Who can you call, text, or e-mail when something happens?

Have you ever attended your sibling's heart doctor appointments? Why or why not?

Did/do your friends know your sibling(s) has LQTS? Do they ask about it?

How did/do you get along with your brother(s)/sister(s) with LQTS?

Did/do you do activities together?

How did/do you get along with your mother?

How did/do you get along with your father?

Has your relationship changed with either/both parent(s) since your sibling(s) was diagnosed with LQTS? If so, how?

Has your relationship changed with your sibling(s) since he/she was diagnosed with LQTS? If so, how?

How do you think your parents treat(ed) your brother(s)/sister(s)?

Were/are you treatment the same or different? If different, provide examples.

Is there anything else you think is important for us to know? If so, please describe here.

Have you been involved in any other LQTS related research projects at the Philadelphia College of Osteopathic Medicine?