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Anxiety in Individuals Affected by Long QT Syndrome as Experienced by Members of an Online User Group

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Philadelphia College of Osteopathic Medicine

Department of Psychology

ANXIETY IN INDIVIDUALS AFFECTED BY LONG QT SYNDROME AS
EXPERIENCED BY MEMBERS OF AN ONLINE USER GROUP

By Abigail M. Janney, M.S., M.S.

Submitted in Partial Fulfillment of the Requirements of the Degree of

Doctor of Psychology

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

Dissertation Approval

This is to certify that the thesis presented to us by Abigail Janney
on the 26th day of April, 2011 in partial fulfillment of the requirements for the degree of
Doctor of Psychology, has been examined and is acceptable in both scholarship and
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Abstract

Long QT Syndrome (LQTS) is a cardiac condition that affects approximately 1 in 2,500 persons. This condition has been vastly understudied with regard to psychosocial aspects. LQTS patients have the potential for experiencing life threatening cardiac events and prevention requires significant lifestyle modifications. Although there has not been a significant amount of research on this specific syndrome, other research on chronic illnesses suggests that many individuals experience significant psychosocial aspects after diagnosis. This study specifically examined anxiety within the LQTS population through a qualitative research design. An archived database from a LQTS user group involving approximately 780 members was used to ascertain anxiety related themes. Results of the research provided two distinct periods in which participants expressed anxiety related to the diagnosis. These results are discussed within a cognitive behavioral context to elaborate upon identified themes within this population, including ways in which these responses could become maladaptive. Although the primary goal of this research was to identify specific themes related to anxiety, it also serves as a stepping-stone for future research within the LQTS population.

Table of Contents

Acknowledgements.....	iii
Abstract.....	iv
Chapter 1.....	1
Introduction.....	1
Statement of the problem.....	1
Purpose of the study.....	2
Overview of the literature review.....	3
Chapter 2.....	4
Review of the literature.....	4
Long QT syndrome.....	5
<i>Mechanisms and characteristics of LQTS</i>	5
<i>Diagnostic criterion for LQTS</i>	7
<i>Medical management of LQTS</i>	13
<i>Summary</i>	19
Introduction to chronic disease and illness.....	20
<i>Specific chronic illness underpinning for adolescents and families</i>	25
<i>Summary</i>	31
Theoretical explanations of psychosocial implications of LQTS.....	31
<i>Introduction to stress and adaptation</i>	39
<i>Cognitive models</i>	41
<i>Transactional stress and coping model</i>	47
Overview of anxiety.....	59

<i>Panic disorder with and without agoraphobia</i>	61
<i>Specific and social phobia</i>	61
<i>Obsessive-compulsive disorder</i>	62
<i>Posttraumatic stress disorder and acute stress disorder</i>	62
<i>Generalized anxiety disorder</i>	63
Conclusion	64
Research question	65
Chapter 3.....	63
Methods.....	63
Overview	63
Design and design justification	63
Participants.....	68
Inclusion and exclusion justification	68
Screening procedures determining inclusion and exclusion criteria.....	68
Recruitment.....	69
Measures	69
Procedure	69
Chapter 4.....	72
Results.....	72
Initial treatment of LQTS.....	73
<i>Uncertainty</i>	74
<i>What syndrome</i>	74
<i>Waiting game</i>	75

<i>Unpredictability</i>	77
<i>Big changes, really quickly</i>	77
<i>It's serious, so what do i do now</i>	78
<i>Uncontrollability</i>	79
<i>Learning to walk again</i>	79
<i>On top of everything else</i>	80
Continued treatment of LQTS.....	81
<i>Uncertainty</i>	81
<i>What if's of medical procedures</i>	81
<i>Future concerns</i>	83
<i>Unpredictability</i>	84
<i>Silent striker</i>	84
<i>Parental concerns</i>	85
<i>How many</i>	86
<i>Uncontrollability</i>	87
<i>No voice</i>	88
<i>Support</i>	89
Conclusion.....	89
Chapter 5.....	91
Discussion.....	91
Development of anxiety and implications to LQTS.....	91
Cognitive behavioral model of anxiety.....	92
Biopsychosocial model.....	97

Role of clinical health psychologist	99
Future research	100
Limitations	101
Conclusion	102
References	103

List of Figures

Figure 1. Diagram of initial treatment themes.....112

Figure 2. Diagram of continued treatment themes113

Chapter 1

Introduction

Statement of the problem.

Approximately 12% of individuals in the United States are living with a chronic disease. Similar rates occur across the world's population with an estimated 10% of individuals affected by chronic illness (Martz & Livneh, 2007). Chronic illness is defined as a disease that has a prolonged course in which individuals can sustain normal lifestyles, or as a disease that can be progressive and possibly fatal (Kanney, Hamrin, & Grey, 2003). Chronic illness defines adolescents and their family's planning, actions, and feelings over a significant length of time or throughout their lifetimes (Sieffe-Krenke, 2001). Clearly, an illness of this nature presents multiple obstacles and ramifications for adolescents and their families, potentially leading to emotional and physical difficulties (Major, 2003). Because of the significant impact of having a chronic illness, it is imperative to understand how this illness affects these individuals' functioning within different aspects of their lives.

Long QT Syndrome is a chronic illness, often diagnosed in adolescence. Most individuals, even those affected, are unaware of this condition prior to their diagnosis. It is estimated that 1 in 2,500 births could be diagnosed with this hereditary, cardiac condition. This disease is represented by a prolongation of the QT interval, and an associated risk of life-threatening arrhythmias (Crotti, Celano, Dagradi, & Schwartz, 2008). This syndrome is particularly dangerous because of the common absence of a diagnosis prior to life-threatening symptoms or to the occurrence of a fatal episode. Although this chronic illness poses significant health risks, there has been a lack of psychosocial research about adverse consequences related to the syndrome. It is

suggested that an increased risk of negative psychosocial consequences occurs in the context of chronic illnesses. Anxiety is pervasive in chronic illness populations that require lifestyle modifications and medication adherence. It is estimated that in some chronic illness populations, anxiety occurs in 49% of those directly affected by the illness (Papneja & Manassis, 2006). Similar to other chronic illnesses, individuals affected by LQTS may experience anxiety. It is suggested that this anxiety may be related to different mechanisms of actions and factors, compared with other chronic illnesses.

Purpose of the study.

The purpose of the present study is to examine anxiety experienced by individuals affected with Long QT syndrome (LQTS). Previous chronic illness research indicates the pervasiveness of this illness not only for the individual, but also for those directly involved with the individual. Within the LQTS population, adolescents and their families are particularly important to consider. Research on other chronic illnesses indicates that those diagnosed in adolescence experience significant anxiety-related symptomology as do their family members. One must be cautious in applying these studies to individuals with LQTS because there is little specific psychosocial research in this area, leading to a lack of knowledge and understanding of multiple aspects of this condition. Well-researched chronic illnesses will be utilized as guidelines or as a starting point to conceptualize anxiety-related symptomology in LQTS. Utilizing this knowledge, this study seeks to identify trends and specific factors related to anxiety within this population. It is suggested that this study will provide information about anxiety that has been exacerbated by a diagnosis of LQTS through identification of themes within the data. This will provide a framework for future research aimed at the understanding of implications of anxiety in this population.

Overview of literature review.

The subsequent literature review will provide information about LQTS, including medical implications of the syndrome, medications, prevalence rates, onset, and duration. LQTS will be compared with other chronic illnesses. Various aspects of chronic illness will be discussed, including the different areas of life that are affected by the illness, as well as medical and psychological symptomology. The psychological sequelae of chronic illnesses will be discussed as a comparative tool to understand potential psychosocial ramifications of LQTS. Last, a brief conceptualization of adaptive anxiety verses maladaptive anxiety will be provided.

Chapter 2

Review of the Literature

Chronic disease is a common experience of countless individuals. As advancements in technology increase life expectancy, more individuals have the possibility of experiencing a chronic illness during their lifetimes. It is estimated that 12% of individuals in the United States are living with a chronic disease. Similar rates occur across the world's population, with an estimated 10% of individuals being affected by chronic illness (Livneh & Martz, 2007). An important sub-grouping within these estimates includes adolescents and their families. Families need to be considered within this context because of their roles in adolescents' lives. Chronic diseases are a multidimensional phenomena that affect nearly 10% to 15% of adolescents and their families (Vitulano, 2003).

Long QT syndrome (LQTS) is a chronic disease on which limited psychosocial research has been performed. This disease shares multiple commonalties with many of the chronic diseases that are studied extensively. Many discoveries regarding LQTS have been made in the past 2 decades; however, medical uncertainties persist regarding diagnosis, treatment, and prognosis. Additionally, many psychosocial aspects of this syndrome are still unknown. One understudied aspect is how some individuals affected by this syndrome effectively control illness related stressors, but others develop anxiety. It is very important to have a solid working knowledge of the biological aspects of this syndrome to understand the short and long-term implications of the medical requirements and lifestyle modifications. This knowledge informs understanding of the development of anxiety within this population.

Long QT syndrome.

LQTS is a hereditary cardiac condition characterized by specific cardiac symptomology that can result in devastating consequences, especially if it remains undetected (Zareba & Cygankiewicz, 2008). This condition is an ion channel disease, in which individuals have a structurally normal heart with an electrical abnormality due to mutations in genes that affect ion channel structure or function. LQTS is a notable syndrome because it often leads to sudden death in seemingly healthy individuals (Vetter, 2007). Currently, it is estimated that the prevalence rate of LQTS is 1 in 2,500 persons (Crotti, Celano, Dagradi, & Schwartz, 2008). A majority of cases (approximately 85%) are inherited, whereas the other 15% are sporadic cases that are due to new genetic mutations in the affected individual and are not inherited (Zareba & Cygankiewicz, 2008). Jervell and Lange-Nielsen initially reported this syndrome in a family with prolonged QT intervals, hereditary deafness, and sudden death. Subsequent reports of similar symptomology and findings, but without congenital deafness have led to the recognition and designation of LQTS. The development of a LQTS registry and extensive medical research in this area has resulted in recommended treatment modalities for this syndrome (Goldenberg & Moss, 2008).

Mechanisms and characteristics of LQTS.

LQTS is characterized by a prolonged QT interval that can lead to symptomatic events including syncope, cardiac arrest, or sudden cardiac death (Zareba & Cygankiewicz, 2008). This prolongation of the QT interval is due to mutations in genes that affect the structure and function, most commonly, of the sodium, potassium, and other ion channels. An individual with LQTS has abnormal ion channel currents that lead to prolonged repolarization or recharging of the heart (Goldenberg & Moss, 2008).

Research has identified different presentations and characteristics of this syndrome. Thus, it is essential to understand the variation among the identified forms of LQTS.

More than 10 genes related to LQTS have been identified, with hundreds of mutations in these genes. The most common genes identified in individuals are those responsible for LQT1 and LQT2. It is estimated that approximately 90% of LQTS diagnoses are identified as LQT1 and LQT2. LQT3 accounts for approximately 10% of the population, and the remainder of identified LQTS genes are considered less common (<1-5%) (Zareba & Cygankiewicz, 2008).

The LQT1 mutation is responsible for approximately 40 to 45% of genotyped cases of this syndrome (Zareba, 2006). It has been shown that this mutation leads to decreased potassium channel function, which in turn leads to decreased potassium channel current (Zareba & Cygankiewicz, 2008). LQT2 accounts for 40 to 45% of LQTS cases (Zareba, 2006). It also is suggested that LQT2 operates by the same mechanism of decreased potassium channel function; however, the gene mutation is different from LQT1 (Zareba, 2006). LQT3 is relatively rare and accounts for approximately 10% of identified cases. LQT3 is caused by a late increase in the sodium channel current or gain of function of that channel. The gene variations of this syndrome suggest different electrophysiological mechanisms based on ion channels that lead to differences in presentation, specific types of exacerbating events and treatments (Zareba & Cygankiewicz, 2008).

Based on the varying electrophysiological mechanisms that cause LQTS, symptomatic events are triggered by different stimuli. Cardiac events for LQT1 are more likely to occur during strenuous physical activities, including exercise and sports (Zareba & Cygankiewicz, 2008). Although approximately 86% of individuals with LQT1 have a

cardiac event by the age of 20, this syndrome often goes undetected until the first event occurs (Schwartz et al., 2001). Untreated individuals with LQT2 often have triggering events related to arousal, emotional stress, auditory stimulation, postpartum changes, and drugs (Vetter, 2007). A cardiac event for LQT3 often occurs while an individual is asleep or at rest, with only a few events related to exercise or emotional distress (Zareba & Cygankiewicz, 2008). Individuals with LQT1 have more events, compared with LQT3, but those with the LQT3 have a greater likelihood of experiencing a fatal event (Vetter, 2007).

Gene-specific diagnosis of the type of LQT indicates specific triggers that may precipitate a cardiac event. In many instances, it may prove difficult to modify certain environmental triggers outside of the home. Individuals must also be aware of triggers and be compliant with the proper course of treatment. Unless an individual is diagnosed with LQTS either by having an electrocardiogram (ECG), a symptomatic event, or a diagnosed family member, he or she is unaware of the need to take precautions. Some individuals exposed to triggers remain asymptomatic, but others may experience life-threatening events early in life. Because of the nature of this syndrome, even those who are diagnosed and treated, have a somewhat unpredictable course throughout life, which may contribute to anxiety.

Diagnostic criterion for LQTS.

Schwartz provided diagnostic criteria based on clinical history, family history, and the results of ECGs, devices that measure the electrical activity of the heart. The diagnostic scale provides three different probability classifications for the clinician. Based on these criteria, an individual's risk can be low probability (or less than one point), medium probability (which corresponds to two to three points), or high probability

(which is \geq four points). Those individuals that are considered at the lowest risk level include males with LQT1 and LQT2 that have corrected QT intervals of less than 500 milliseconds (Priori et al., 2003). No prior history of syncope places one in a lower risk level, as well. (Goldenberg & Moss, 2008). The next level, intermediate risk, has an estimated 30% to 49% probability of a cardiac event before age 40 years if the individuals are not on medication. Females with LQT2 and LQT3 and males with LQT3 even with QT intervals less than 500 milliseconds are part of the composition of this group. Females with LQT3 and individuals with QT intervals greater than 500 milliseconds are also included within this risk level (Priori et al., 2003). These individuals may also have a history of prior syncope (Goldenberg & Moss, 2008). The highest risk suggests a 50% probability of experiencing a cardiac event before the age of 40. Those individuals with LQT1 or LQT2 or males with LQT3 who have QT intervals of greater than 500 milliseconds compose this group (Priori et al., 2003).

In assessing an individual for LQTS, the first priority is to obtain an extensive clinical history to identify previous symptoms related to LQTS (Vetter, 2007). A major component of the clinical history is to identify prior episodes of syncope. Recent episodes of syncope are not only a symptom of LQTS, but also increase the probability of a subsequent cardiac arrest or death (Zareba & Cygankiewicz, 2008). It is important to identify when the episode of syncope occurred and whether or not it was concurrent with a stressful event, either emotional or physical. Another important association with LQTS is congenital deafness, a reflection of another LQTS type known as Jervell and Lange-Nielsen syndrome. This syndrome is a recessive variant of LQTS (Crotti, Celano, Dagradi, & Schwartz, 2008). Children inherit this syndrome from both parents, resulting in gene mutations on both alleles. LQTS without hearing deficit, is inherited in a

dominant fashion with only one parent having the specific mutation and thus, only one allele involved. The genes that are responsible for JLN regulate the potassium channels in both the heart and the ear (Vincent, 2005).

After obtaining a complete personal history, the clinician needs to obtain a complete family history. The family history may highlight cardiac events that include syncope, sudden death including sudden infant death syndrome, drowning, or unexplained motor vehicle accidents. This review should include both extended and immediate family members. Gathering all pertinent information is essential in order to make a correct diagnosis of LQTS. Additionally, one should include other historical factors when acquiring this data, such as congenital deafness in family members (Goldenberg & Moss, 2008).

The clinical course of this syndrome can vary depending on genotype and environmental factors, age and gender, historical events, and therapy (Goldenberg & Moss, 2008). An important area of information regarding the clinical course of the syndrome relates to genotype. Those individuals with LQT1 and LQT2 are more frequently prone to cardiac events than those with other mutations especially if they have QT intervals greater than 500 milliseconds. However, individuals with LQT3 are at the highest risk for fatal cardiac events, when compared with LQT1 and LQT2 (Zareba & Cygankiewicz, 2008). As previously noted, the clinical course of this syndrome is highly dependent on the interaction between specific gene mutations and environmental triggers (Goldenberg & Moss, 2008). Awareness and understanding of the probability of an event based on the genotype and environmental factors are important to determine the prognosis and treatment plan, which then affect the clinical course and quality of life.

After puberty, females have longer QT intervals than males. Moreover, clinical research suggests a higher incidence of this syndrome within the female population (Zareba & Cygankiewicz, 2008). There are differences between genders with regard to cardiac events as well. Males are at higher risk than females for a cardiac event, until approximately 12 years of age. Speculated to relate to hormonal changes, females develop greater risk for a cardiac event after puberty. Women are also at higher risk for cardiac events during the post-partum period and menstruation. This increased risk may be related to elevated hormonal levels that influence ion channels, or may be due to increased sympathetic activity (Goldenberg & Moss, 2008).

Gender differences are found in the different subtypes of LQTS. Before puberty, male children with LQT1 have a higher risk than same aged females. This pattern reverses in adulthood when adult females with LQT1 develop a higher risk for cardiac events than adult males. Children with LQT2 have a lower risk for cardiac events with less differentiation between genders at this age. In adulthood, females with LQT2 have a significantly higher risk than males. Interestingly, although females are at higher risk for an increased number of cardiac events, males with LQT1 and LQT2 remain at higher risk for lethal cardiac events throughout their lifespan. There is no clear pattern in those with LQT3 concerning gender (Zareba et al., 2003).

Clinical information from electrocardiograms can help to characterize those with LQTS. QT intervals are an important diagnostic factor used to stratify an individual's risk for cardiac events. Individuals with corrected QT intervals greater than 500 milliseconds are suggested to have an increased risk of a cardiac event (Goldenberg & Moss, 2008). It is suggested that each 10-millisecond corrected QT interval increment increases the risk of a cardiac event by 5% (Zareba & Cygankiewicz, 2008). Follow-up

ECGs are recommended to evaluate the likelihood of future cardiac events (Goldenberg & Moss, 2008).

The presence of bradycardia is another important risk factor. Individuals with a history of untreated bradycardia were twice as likely to experience a cardiac event in comparison with individuals with normal heart rates. Individuals with a prior syncope event have a higher probability of future cardiac events (Zareba & Cygankiewicz, 2008). Previous history of syncope should serve as a warning to provide further evaluation and treatment in individuals with LQTS.

The ECG can identify specific patterns that correspond with LQTS subtypes (Zareba & Cygankiewicz, 2008). The ECG shows the length of time between the start of the Q wave and the end of the T wave, which represents time for the heart to depolarize (discharge) and repolarize (recharge). The length of time between these two, the onset of depolarization and the end of repolarization, is known as the QT interval, and a longer interval than normal represents an electrical instability in the heart's rhythm, increasing the risk of a cardiac event as the QT interval become increasingly longer (Zareba & Cygankiewicz, 2008). Published reports indicate that a QT interval of more than 500 milliseconds confers a two to three fold increased risk of cardiac arrest, sudden death, or syncope as compared with those individuals with normal QT intervals (Zareba & Cygankiewicz, 2008). QT intervals can be normal in individuals with LQTS and can be slightly elevated in those without the syndrome, resulting in an overlap of QT intervals in affected and unaffected individuals. Variations in QT intervals and in characteristics of the syndrome are the result of differences in gene expression and reentrance. The most malignant arrhythmia or abnormal rhythm associated with LQTS is a malignant type of ventricular tachycardia known as torsades de pointes. This arrhythmia often results in a

sudden loss of consciousness and can lead to death if not addressed promptly. In the visual representation of torsades de pointes, it appears as though the QRS axis is performing a continuous twist around the baseline. Torsades de pointes can spontaneously correct itself or it deteriorates to ventricular fibrillation that can result in death if not corrected. The self-correcting nature of these cardiac patterns explains the presence of multiple syncopal events before the occurrence of a fatal event (Wehrens, Vos, Doevendans, & Wellens, 2002).

ECGs can identify other cardiac abnormalities that occur in this syndrome, including T-wave abnormalities and bradycardia (Zareba & Cygankiewicz, 2008). T-waves are inscribed during cardiac repolarization and abnormalities represent abnormal cardiac repolarization, a potential cause of significant concern. There are several variations of T-wave abnormalities that can occur in an individual with LQTS. It is important to consider the fact that abnormalities can occur in a healthy individual, but occur more commonly in those who have this syndrome. Those individuals with LQTS who have identified specific T-wave abnormalities are more prone to cardiac arrest and syncope. Interestingly, there are notable differences between T-wave morphology and patterns among the different subtypes of LQTS. LQT1 mutations often have a significantly longer duration of the T wave, referred to as broad based T waves. Those individuals with LQT2 have small or notched T waves. T waves associated with LQT3 can reveal a late onset of the T wave with a long flat ST segment (Zareba, 2006). ECGs also indicate when bradycardia, a lower than normal resting heart rate, is present. Individuals with LQTS may have low heart rates, both during rest and with exercise (Crotti et al., 2008).

Personal and family history and clinical tests, such as ECGs, do not always provide clear identification of this syndrome. Genetic testing provides additional clinical evidence about the individual and family members. Genetic testing is helpful to determine a definitive diagnosis and to identify subtypes to help determine prognosis and treatment regimens. Genetic testing helps to decrease the ambiguity of the diagnosis of this syndrome by providing concrete evidence, and also serving as an outstanding research tool to increase clarity. When the family mutation is known, genetic testing is extremely helpful determine if LQTS is present or not present in other family members. Large scale genetic testing has not been evaluated for those individuals who present with borderline QT intervals, suspicious symptoms, and no family history (Roden, 2008).

This type of testing is available only at a few clinical laboratories, is quite expensive, and is often refused coverage by insurance companies. Seventy five percent of those with LQTS can be identified by genetic testing at the present time. The other 25% of the causative genes are yet to be identified. This suggests that a negative genetic test does not warrant the removal of the diagnosis unless the family gene is known and the family member tested does not carry that gene mutation. These results suggest a less than complete science, for which the testing is extremely costly. However, if a patient is positively identified, this provides impetus for accurate clinical decisions, especially for potential family members who are asymptomatic.

Medical management of LQTS.

The medical treatment of LQTS has made significant progress through the years because a more comprehensive understanding has been provided through research and published reports of individuals who have this syndrome. The treatment of LQTS is complicated, based on the various subtypes and levels of risk and the need for careful

consideration of clinical history. There is general agreement regarding symptomatic patients and their need for treatment, although treatment regimens may differ, especially with regard to the use of defibrillators for individuals who have not had life-threatening events or arrhythmias. Some dissent remains regarding the treatment of asymptomatic patients (Schwartz, 1997).

Schwartz et al. (1997) provided a more systematic understanding of the treatment of asymptomatic and symptomatic individuals. High-risk subtypes that require prompt and aggressive treatment are outlined. The first high risk group includes those with congenital deafness, associated with recessively inherited LQTS, who experience a high risk of cardiac events. Second, prompt treatment is recommended in neonates and infants because of increased risk during the first few months of life. This report recommended treatment when there have been sudden deaths of any siblings in the past because of emotional stress in the environment, but not all would agree with this suggestion. Others at high risk who warrant immediate treatment include those with T wave alternans on ECG, a severe repolarization abnormality, because this provides evidence of increased electrical instability. At particularly high risk are those individuals who have a QT interval of greater than 600 milliseconds, because they are prone to be more symptomatic and experience life-threatening events (Schwartz, 1997). Currently, the treatment modalities for LQTS aim to decrease arrhythmic events, lessening adrenergic response, and prevent sudden death. Overall, these goals aim to prevent fatal ventricular tachycardia, torsades de pointes, and ventricular fibrillation (Zareba & Cygankiewicz, 2008).

The first line of treatment for LQTS continues to be the utilization of beta-blockers. Propranolol is one of the most widely used drug regimens within this

population. This beta blocker is well tolerated in chronic therapy and easily crosses the blood-brain barrier. The main disadvantage of beta-blockers is the need for multiple, daily administrations, and it has been suggested that nadolol or other longer acting preparations be used to reduce the number of administrations. Compliance is a major factor in consideration of treatment modalities, and a lack thereof can have serious consequences for individuals with LQTS (Crotti et al., 2008). Medical recommendations suggest the use of these drugs for all high and moderate risk individuals, as well as for most low risk patients (Goldenberg & Moss, 2008). It has been suggested that in high-risk adolescents, the use of beta-blockers leads to a 64% decrease in the probability of a cardiac event (Zareba & Cygankiewicz, 2008). Results from other studies continue to confirm the overall effectiveness of beta-blockers for high-risk individuals (Goldenberg & Moss, 2008). Similar to other components of LQTS, the type of mutation should be considered in order to determine the likelihood of effectiveness of beta blockers for an individual. Beta blockers have proven to be extremely effective for LQT1; it is somewhat less effective for the LQT2 mutation, however, but still a recommended treatment modality. Individuals with LQT3 still have an elevated risk of cardiac events when on a beta blocker regimen although it is effective in 49% of individuals, but it is recommended that additional therapies are considered, especially in high risk or symptomatic individuals. In addition, those individuals with Jervell and Lange-Nielsen syndrome may require consideration of alternate therapies (Crotti et al., 2008).

The mechanism of action of a beta blocker is related to the role of adrenergic stimuli in cardiac events. Beta blockers inhibit or block hormones and neurotransmitters secreted during times of emotional arousal and physical activity that lead to cardiac events. Some individuals remain at high risk even after the introduction of beta blockers;

however, research has identified a decrease in life-threatening events in high-risk individuals (Goldenberg & Moss, 2008). The continuation of symptoms even after treatment with beta blockers suggests the need for modification of treatment that includes the addition of an implantable cardioverter-defibrillator (ICD) or possible left cardiac sympathetic denervation (Zareba & Cygankiewicz, 2008).

The second form of treatment for LQTS is an implantable cardioverter defibrillator (ICDs). Individuals with repeated syncopal events while on a beta blocker regimen, and those who are considered high risk and have survived previous cardiac arrests are prime candidates for this form of treatment. Other patients that may be considered for this form of treatment are those individuals with a previous episode of syncope with more than 500 milliseconds QT interval and who are receiving beta blockers. As with any treatment modality, there are potential risks and benefits that should be considered. Some drawbacks of ICDs include continual maintenance and replacement of the device, pain and anxiety associated with inappropriate shocks, lead complications, and other possible malfunctions. This continual maintenance and requirement for new devices occur multiple times during an individual's lifetime, even without any complications, because the batteries need to be replaced. Inappropriate shocks are another significant disadvantage of this device. These inappropriate shocks and the pain experienced with each shock can cause significant emotional distress (Crotti et al., 2008). These problems may make an individual or family hesitant to select this treatment option. Thus, the treatment team as well as the family should consider and discuss the risks and benefits extensively, as is the case with all treatment modalities. However, even with all the potential disadvantages, ICD treatment is life saving for high-

risk individuals. In some studies, there was a 16% to 1% decrease in cardiac events in individuals with ICDs (Goldenberg & Moss, 2008).

Another type of treatment modality that is used for LQTS, and often in conjunction with beta blocker treatment, is cardiac pacing, especially in very young children or in those with very slow heart rates. Cardiac pacing is often used with individuals with atrioventricular block, which is a result of conduction disruption between the atria (upper heart chambers) and the ventricles (lower heart chambers). Cardiac pacing is also used for individuals prone to bradycardia most commonly seen in LQT3. Other individuals without these specific characteristics may also benefit from cardiac pacing (Schwartz, 1997).

Another treatment option for this syndrome is left cardiac sympathetic denervation (LCSD). Individuals who undergo this surgical option usually are considered at high risk, and continue to experience syncopal events after other treatment modalities have been introduced (Goldenberg & Moss, 2008). In a sample of 147 high-risk individuals who have undergone LCSD in the past 35 years, this treatment modality appeared efficacious. Of these specific individuals, 99% were symptomatic and had an extremely long baseline QT interval prior to this treatment. These individuals had been previously treated with beta blockers; almost half of these still had a cardiac arrest and 75% still had continual syncopal episodes. After continued treatment with beta blockers and LCSD there was an overall decrease in QT interval length and a 95% decrease in shock count, as well as an overall decrease in mortality rates in this high-risk group (Crotti et al., 2008).

Lifestyle modification should be considered as a form of treatment and prevention of life-threatening events. Consideration of the type of mutation is essential when

considering environmental and lifestyle modifications (Zareba & Cygankiewicz, 2008). Although exercise limitation is recommended for all individuals with LQTS, those with LQT1 should have, modifications related to strenuous physical exertion activities, because they are the most susceptible to the adverse effects of exercise. In most instances, these individuals are advised to avoid physical activity completely, especially if they have a history of syncope or other cardiac events. Another strong indicator for limitation of physical activities is the presence of an ICD or pacemaker related to the potential damage to the unit by contact sports. It is unrealistic to avoid engagement in all physical activities completely. Therefore, it is useful to consider recreational sports rather than competitive ones. Competitive sports often are considered off-limits for individuals with this syndrome. Competitive sports are viewed as those that require systematic training to achieve a physical goal, and also involve the pressure to excel while reaching that goal. In comparison, recreational sports provide a range of physical exertion that an individual does not pursue on a consistent basis in aspiration of a higher end goal (Maron et al., 2006).

A list of activities has been developed that considers multiple variables such as level of exertion and environment to help clinicians advise families on appropriate activities, based on history and determined risk level. For instance, activities such as swimming or ice hockey, which expose a person to harsh environmental conditions that alter body chemistry, are ill advised because of the added physical stress involved. This list was also developed in recognition of the overall quality of life that exercise brings to someone's life, both physically and psychologically. Individuals with LQT2 are expected to modify their environments and lifestyles by considering events or stimuli that may induce emotional arousal or unexpected noises. These individuals are advised to

remove anything that may create loud noises such as alarm clocks or telephones from their environments. Measures should be taken to minimize stimuli (such as starter guns) that are likely to startle the individual (Maron et al., 2004). Competitive sports are generally not recommended.

Common to all mutations, prevention of cardiac events is the goal. Thus, compliance with medical regimens is essential because of the life-threatening nature of this syndrome. Another necessary precaution is awareness of drugs that may alter QT intervals and other body mechanisms that can induce cardiac events. Last, a state of preparedness is also recommended, so that having an automatic external defibrillator (AED) helps negotiate times when other medical modalities do not prevent an event from occurring (Zareba & Cygankiewicz, 2008).

Summary.

Chronic illness is a multifactor phenomenon that presents multiple obstacles throughout its course. Many chronic illnesses have been researched and well documented by clinicians over a significant length of time. All chronic illnesses present challenges; however, LQTS introduces components that are different from other illnesses. A significant factor unique to LQTS is the lack of clear identification of specific treatment modalities for the syndrome. Studies have provided evidence for treatment. However, these outlines are based on prototypical cases that a clinician has to incorporate into a multifactor conceptualization of an individual, based on clinical presentation to individualize the treatment plan. Another critical factor related to this syndrome is the life-threatening nature of the disease. This syndrome presents multiple obstacles for a clinician and individual to overcome through medical regimens, lifestyle control, and medications that seem appropriate for the specific individual. Incorporating these

modifications into one's lifestyle can prove to be overwhelming, and the lack of certainty that comes with the diagnosis may greatly affect one's psychological well-being.

The ambiguity that is present with this syndrome is not common with better understood chronic illnesses. Many chronic illnesses share similar aspects such as uncertainty about treatment, significant lifestyle modifications, or demanding medical routines; however, they generally have definite treatment guidelines or are better understood by the medical community. Asthma and diabetes are chronic illnesses that are well understood by the medical community, and some of the lifestyle changes and medical regimens used for these diseases can be used as models for development of treatment plans specific to LQTS.

Introduction to chronic disease and illness.

Chronic disease refers to the medical model of understanding diseases processes. Chronic diseases often do not have a single cause, a specific onset, or a consistent set of symptoms. The definition of chronic disease also indicates a certain level of incurability (Sperry, 2006). Chronic diseases are often prolonged, lasting 6 months or more with little change or progression in the disease (Barlow & Ellard, 2006). The typical course of chronic disease is often highlighted by periods of exacerbation and remission, and possible degeneration (Sperry, 2006). Chronic disease may limit age appropriate functions, cause disfigurement, require medication and dependence on medical routines, increase need for medical care, or for continued medical treatments (Major, 2003). Chronic disease can also be defined, based on three dimensions, with at least one usually evident in an individual with a chronic illness. These are functional limitations, dependence on compensatory mechanisms, and service utilization (Burns, Sadof, & Kamat, 2006).

The first dimension relates to functional limitations based on disease-specific symptomology. For instance, an individual with LQTS may have limitations placed on his or her physical activity. The second dimension relates to dependence on compensatory mechanisms such as dietary limitations, medications, and equipment (Burns et al., 2006). Individuals afflicted with a chronic illness must endure continuous management of a disease with no cure (Hill-Briggs, 2003). Specific to mutation type, individuals affected by LQTS are dependent on these mechanisms. For instance, those with LQT1 have certain physical restrictions and those with LQT3 require specific dietary modifications. The last dimension related to chronic illness is service utilization. Individuals with chronic illness may require services beyond routine medical care including services such as physical therapy or special education accommodations (Burns, Sadof, & Kamat, 2006). Individuals affected by LQTS have frequent medical appointments to ensure that treatment regimens are effective. Certain individuals may also require environmental modifications based on specific mutations.

Chronic illness refers to the subjective experience of chronic disease (Sperry, 2006). This subjective experience is important for the understanding of anxiety within this population, because of varying factors that influence the illness experience. The chronic illness experience is much different from a purely medical model. This experience is influenced by multiple factors including: personal factors, culture, relationships and social support, socioeconomics, activities, environment, and personal goals of the individual. Personal factors interact significantly with the overall disease process. An individual's ability to adapt to an illness is influenced by his or her level of tolerance towards symptoms, general coping ability, and past experiences. Another important personal factor that needs to be considered is functional capacity. Functional

capacity relates directly to an individual's ability to manage disease related circumstances. An individual that is older may become more seriously impaired in comparison with a younger adult because of the syndrome. This capacity can also relate to the adolescent population; this will be further discussed as a subsection of the chronic illness population (Falvo, 2009). Personal factors relating to LQTS and anxiety within this population are currently unknown. Another consideration is that the interaction of personal factors is highly dependent on the individual and therefore, conceptualized accordingly.

Relationships and social support are major factors affecting those with chronic illnesses. These factors have a direct influence on those afflicted with the illness. These relationships can include significant others and members of the medical community. These relationships are reciprocal; not only does an individual's reaction to an illness influence others, but others' reactions influence the individual's reactions. These relationships should also be considered within a wider context of culture and socioeconomic status (Falvo, 2009). Individuals affected by LQTS are confronted with numerous changes in their lives. These changes are highly suggestive of the need for support throughout the process. Perhaps lack of support or lack of proper relationships leads to anxiety within this population, but this is not known.

Environmental factors encompass three areas: physical, social, and political. Environmental factors should be considered in context of the level of adaptation needed, based on specific chronic diseases (Falvo, 2009). Individuals affected by LQTS have varying factors to consider, based on specific mutations and environmental contexts. An individual with LQT1 must be mindful of physically demanding activities, whereas individuals with LQT2 must be more aware of emotion-arousing stimuli (Zareba &

Cygankiewicz, 2008). Environmental factors should be considered not only as they relate to specific mutations, but also as they relate to flexibility in modification. An individual affected by this syndrome, who has interests or activities that do not correspond with medical recommendations may be more likely to experience anxiety. Environmental variables can also be considered in relation to support and relationships. These relationships should be considered in conjunction not only with common environments, such as academic or home settings, but also within medical and other illness related environments. It is suggested that anxiety may be directly related to the interaction between LQTS and specific environmental variables.

The specific goals of an individual should also be considered within the context of chronic illness (Falvo, 2009). An individual affected by LQTS may or may not have goals that are affected by the syndrome. An individual that has goals related to physical activity may be more highly affected than those who have academic goals. These goals also interact not only with the specific physiology of the syndrome, but also with other personal factors. Anxiety may be related to the loss of control related to goals.

The numerous factors that are influential within chronic illness speak to the need for a specific understanding of the disease process related to individual illness factors. Within the LQTS population, little research has been done to truly understand the illness experience; this may influence the development of poor adaptation and anxiety related responses. Based on other similar chronic illness research, there can be speculation about those factors that lead to anxiety in these individuals. However, it becomes crucial to develop an understanding based on the specific biological mechanisms and subjective experiences related to this syndrome. These subjective chronic illness experiences may

be case-specific. However, it is hypothesized that certain themes or triggers within these variables lead to anxiety responses in certain individuals.

To organize these factors into a systematic order, chronic illness can also be conceptualized within a phase model. Fennel (2003) developed a phase model of chronic illness that allows for both progression and regression through the course of chronic illness. The first phase, crisis, occurs during the initial diagnosis, and other crises occur throughout the course of the illness. Individuals cope with this information through medical treatment, support, or negative behavior, such as substance abuse. Families react to the diagnosis with disbelief, rejection, or horror. The main goal of this phase is to deal directly with the symptomology of the illness (Sperry, 2006). An individual affected by LQTS may require implantable devices and medication routines. These modifications would be handled during this phase. These modifications would also be influenced by other illness variables, such as personal factors or support systems. Anxiety-related responses could develop during this phase because of the presentation of chronic disease.

The second phase, stabilization, occurs at the time when an individual becomes more familiar with the illness. Individuals often make attempts to regain a sense of normalcy in varying environments during this phase. The goal of this phase is to acclimate to lifestyle modifications attributed to the chronic illness (Sperry, 2006). Individuals affected by LQTS may require altering the physical environments or daily activities occurring during this phase. Chronic illness factors would also be influential during this period because of the adaptability of others and environmental contexts. The consideration of personal goals during this phase is critical because of the way in which they are affected by lifestyle and medication requirements. It is suggested that there may be recognition of the discrepancy between the individual's previous lifestyle and the

altered one after this phase. It can be hypothesized that this evokes a sense of minimal control and the possibility of anxiety-related responses.

The third phase, resolution, occurs at the time when the individual and others have accepted the diagnosis. With this acceptance comes an understanding that their sense of self is altered, based on the specific chronic disease. This acceptance allows the person to develop a new sense of self that incorporates the disease into his or her life (Sperry, 2006). An individual affected by LQTS may be present in this phase as one that has accepted the loss of physical activities or acknowledges the need for continuous management of LQTS. It is hypothesized that by the time an individual reaches this phase of illness, anxiety-related responses have been resolved in previous stages.

The last phase, integration, implies that an individual is able to view his or her illness as only one aspect of his or her life. It should be noted that progressing through this cycle is not a linear process. In addition, individuals may not reach all the different phases and may continuously endure crises that destabilize their emotional states and lead to regression. It has been suggested that these continual setbacks happen continually unless an intervention is implemented (Sperry, 2006). At the current time, proper interventions are not known for the LQTS population. The current research aims to gain insight into these specific factors that lead to continued anxiety responses. It is hypothesized that individuals affected by LQTS that reach this phase of integration do not experience anxiety responses.

Specific chronic illness underpinning for adolescents and families.

Chronic illness is a complex entity that requires a comprehensive understanding of variables that influence the overall process. This understanding becomes more complex when an adolescent has a chronic illness and family dynamics influence the

illness experience. The multidimensionality of chronic illness has already been discussed in relation to factors that are influential across the lifespan. It is also essential to put into context how chronic illness may affect adolescents and their families in alternative ways. Chronic illness presents a number of obstacles to overcome in combination with normal developmental milestones and common daily stressors. Adolescents with chronic illness must maintain medical regimens; undergo medical procedures, multiple hospitalizations, and possible peer rejection because of lack of interaction and physical limitations (Hampel, Rudolph, Stachow, Lab-Lantzsch, & Petermann, 2005). In combination with these medical obstacles, as the adolescent grows there are new challenges in relation to autonomy, physical development, identity formation, cognitive functioning, intimacy, and achievement, which continually cause new dilemmas and challenges (Englund, Hartman, & Segesten, 2006).

Adolescents and their families encounter multiple obstacles throughout the course of a chronic illness. When conceptualizing an individual and his or her family, it is imperative to recognize the multiple life systems that the illness affects for adolescents and their families (Olsson, Boyce, Toumbourou, & Sawyer, 2005). A typical developmental course for an adolescent is significantly altered by chronic illness. An adolescent develops a recognized need for autonomy from the family and a more intense focus on peer groups. As an adolescent develops a sense of autonomy, his or her competence will increase in relationship to his or her ability to manage social relationships, academics, and other important areas (Vitulano, 2003). However, chronic illness limits this autonomy and increases an adolescent's dependence on family members. This dependence alters family dynamics and developmental stages, and possibly decreases the emergence of supportive peer groups (Olsson, et al., 2005). This

decreased autonomy may prevent an adolescent from interacting in a developmentally appropriate fashion, and may lead to overall feelings of decreased self-worth, attributed to perceived low competence. This disengagement may also contribute to conflicted family dynamics, especially if the adolescent blames one's parents for his or her limited lifestyle (Vitulano, 2003). Many families may have difficulty adapting to the changing needs of the child. As the adolescent assumes more responsibility, parents may have difficulty relinquishing this control for various reasons. The lack of control felt both by the parent and by the child may also contribute to increased conflict and blame that could serve only to exacerbate symptomology (Beveridge, Berg, Wiebe, & Palmer, 2006).

Although each chronic illness affects different areas of functioning and requires attention to varying aspects, many alter two of the biggest realms for adolescents: academics and extracurricular activities. The academic setting provides the opportunity for adolescents to develop peer relationships, learn adaptive coping mechanisms, and learn academic material. The ability of the adolescent to perform within the school setting depends not only on parental constraints, but also on the level of medical attention required by the chronic illness. Although the academic setting provides the opportunity for the adolescent to increase his or her competence level to a higher degree, it can also have devastating effects on him or her if removed from this setting for significant periods of time due to medical needs. Parents must also learn to adapt to the school environment, recognize the level of protection required, and learn how to help their child adapt to the environment with a chronic illness (Vitulano, 2003).

Extracurricular activities, including sports participation, are an essential component of the academic setting. It allows adolescents to develop skills of commitment and cooperation while developing relationships with peers. Adolescents

with chronic illness are often limited by medical restrictions, lack of time, or limited strength and endurance (Vitulano, 2003). These physical restrictions place burdens on parents to meet needs in an alternative way and overcome barriers, including financial or time considerations that accompany a chronic illness. Similar to other aspects of chronic illness, this places a great amount of pressure on parents to find resources that will help their child maintain a normal developmental course.

Accurate conceptualization of how chronic illness affects an individual and his or her family is essential in order to provide appropriate treatment and to ensure healthy development and growth. On a daily basis, adolescents encounter new trials to test their characters, and both physical and emotional changes that are at times difficult to understand and adapt to in their daily lives. Chronic illness is an additional factor that can further complicate an adolescent's life. Although adolescents encounter direct physical symptomology because of the demands placed on other family members, the entire family is increasingly susceptible to mental and physical difficulties (Major, 2003). Barlow and Ellard (2006) provided results from a sample of 106 children with chronic illness, suggesting that 64% had mild psychological dysfunction and 50% had psychiatric diagnoses (Barlow & Ellard, 2006). Papneja and Manassis (2006) reported that up to 40% of adolescents with asthma had an anxiety disorder (Papneja & Manassis, 2006).

These psychosocial consequences exist not only within the individual afflicted with the disease, but also directly affect family members. These adjustments in family dynamics and routines place a significant burden on members of the family, which increases the opportunity for emotional difficulties. It has been suggested that these negative consequences for parents are exacerbated by various events related to chronic illness. It has also been suggested that the four most stressful periods of chronic illness

are the initial diagnosis, continuous health care needs of the child, developmental milestones, and illness complications and hospitalizations (George & Vickers, 2006). Anecdotal evidence has indicated a wide range of emotional consequences during these illness periods. These experiences include a sense of shock and disbelief, feelings of guilt, decreased self-worth, and lower self-confidence. These experiences account for a wide range of emotional disturbance. Some common emotional parental experiences indicate a sense of uncertainty and helplessness toward their child's chronic illness. These continual feelings of uncertainty and lack of control over chronic illness increase the possibility of psychological distress (George & Vickers, 2006).

Lack of certainty and control are two crucial factors to understanding how a family copes with chronic disease. Chronic illness and anxiety seem to be interrelated to a great extent, based on previous research completed on various chronic illnesses. These factors exacerbate family anxiety that impacts immediate family functioning and social, emotional and financial areas (Gerhardt et al., 2003). Maikranz, Steele, Dreyer, Startmnet, and Bovaird (2007) found results similar to other research suggesting that decreased uncertainty decreases psychosocial distress and increased adaptive coping mechanisms (Maikranz, Steele, Dreyer, Stratman, & Bovarid, 2007). Parental uncertainty has been suggested to correlate highly with anxiety and depression (Mullins et al., 2007).

An understanding of these factors that tend to exacerbate family stress or anxiety is imperative in gaining a sense of the factors that lead to psychosocial consequences, namely anxiety. Research has indicated numerous specific consequences of increased family anxiety including decreased treatment compliance, direct physiological responses, and decreased emotional control (Major, 2003). An understanding of these cycles can

help professionals intervene, so that the needs of the chronically ill individual are met and the physical and mental health of the parents are attended to in an appropriate fashion. The attention to these functional components of the illness provides the opportunity for parents to meet the demands of the illness, but also attend to and meet each other's needs, including care of siblings, career, or basic self-care (Major, 2003).

It is imperative to recognize parents' perceptions of the illness experience.

Parents provide emotional and physical support to their children throughout the chronic illness experience. The parents' level of responsibility and commitment to chronic illness tasks leads to perceptions of shared illness experiences and events. The high level of commitment that is required of parents leads to a perception of the adolescent's chronic illness as a shared entity. Beveridge, Berg, Wiebe, and Palmer (2006) found that mothers and adolescents perceive their chronic illness as a shared entity. Interestingly, mothers and adolescents did not have congruent views on the most stressful aspects of the chronic illness. In relation to care of a diabetic child, mothers often found diet regimens, poor medical regimens, and interpersonal conflict with their adolescent to be the most stressful aspects of the illness. Adolescents reported metabolic control to be the most stressful event (Beveridge et al., 2006).

The concept of chronic illness as a shared entity provides significant information about the dynamic shifts caused by chronic illness. First, it suggests how individuals' lives are affected by chronic illness. The level of lifestyle modification that is required by those afflicted by a chronic illness is extensive and life altering. Second, this research indicates that although illness management may be viewed as a shared entity, the causes of stress should be considered, based on the role of the individual. This consideration allows an understanding of specific concerns of the individual, based on his or her

responsibilities in relation to the chronic illness and an understanding about how an individual copes with illness related stressors. The understanding of individual stressors also leads to an understanding about how anxiety develops within individuals (Beveridge et al., 2006).

Summary.

Chronic illness presents numerous challenges that require adaptation. These challenges are not only endured by the individual with the illness, but also by family members and others who have a relationship with the individual. The challenges of chronic illness are not due to a concrete set of variables; rather, they are based on specific disease mechanisms and illness factors specific to each individual. For an individual to adapt to these lifestyle modifications successfully there has to be an understanding of those specific factors and responses that led to adaptation. An understanding of coping and adaptation from a theoretical standpoint provides a framework to help understand those variables, which are influential in the adaptation process. Through this understanding, one can start to develop an understanding of anxiety responses within this population.

Theoretical explanations of psychosocial implications of LQTS.

LQTS presents multiple concerns to those affected by this syndrome. LQTS presents life altering changes and uncertainties about the proper course of action in many circumstances for individuals affected by this syndrome. There is limited knowledge about how much these uncertainties influence the psychosocial aspects of the syndrome, but it has been suggested that they are a major aspect of adjustment to any chronic illness experience. Initially, a review of the limited research on LQTS is critical because of the differences found when compared with more highly researched chronic diseases. After

this review, based on this literature and the generalizability of aspects of other chronic diseases, theoretical models can be applied to gain a proposed conceptualization of psychosocial consequences.

Farnsworth, Fosyth, Haglund, and Ackerman (2006) completed initial qualitative research with 31 parents of adolescents with LQTS to describe the experience of families affected by LQTS and the related psychosocial aspects. Research has suggested three areas of concern that are pervasive in the parents of children with LQTS: fear of death, quality of life, and uncertainty. Fear of death was the first psychosocial implication expressed by the majority of parents. Parents described images of the fear of discovering that their child has had a near fatal event and how these fears have played into their desire to protect their child. Many parents described significant lifestyle alterations that helped to decrease the fear of death caused by this syndrome. Many parents described frequently checking on their sleeping children or leaving phone messages while their children are doing activities without them. Other attempts to decrease fear were in educating schools and various organizations, and having an emergency plan established for their child. Parents also described a developmental process of teaching their child about their health and how to become more autonomous through management of their own health (Farnsworth, Fosyth, Haglund, & Ackerman, 2006).

The second psychosocial implication of LQTS was quality of life. Unlike many chronic illnesses, triggers that lead to life threatening cardiac events are often present in common daily environments. These triggers require significant modification of lifestyles and awareness of these triggers. Decisions about treatment modalities must also be made according to specific mutations, side effects, maintenance, and complications of specific medications. Each treatment option has potential advantages and disadvantages that may

affect individuals' quality of life. Parents described making decisions about life saving treatment options that have significant disadvantages, such as long-term maintenance or extreme tiredness (Farnsworth et al., 2006).

Parents indicate that the hardest aspect related to quality of life is the lifestyle modifications. Parents indicated that making difficult decisions about limiting their children's activities created significant stress and placed burdens upon them in the decision making process. Parents described making informed decisions based on the potential consequences and limits, yet trying not to be overly protective of their children. Parents are forced to withdraw their children from rewarding activities, such as sports or other activities that present too many risks, which may evoke conflicts within their household. Parents who coped well with these decisions suggested that being better informed about decisions was helpful, rather than making decisions based on fear. It has also been suggested that families that have had a greater opportunity to adapt to these modifications cope better than families who are diagnosed later in life, or whose life previously revolved around activities that are not significantly restricted (Farnsworth et al., 2006).

Parents must learn how to manage this syndrome effectively and still maintain quality of life. It has been suggested that those parents that are educated about this syndrome are more capable of maintaining quality of life, compared with individuals that lack an understanding or that have other exacerbating factors. Considerations of other factors that may influence quality of life are also crucial to conceptualization in order to understand further the possible psychosocial consequences of this syndrome. Families whose lifestyles had previously involved music and arts, primarily, were less seriously affected than highly athletic families, and may not have to alter their lifestyles or have to

adapt their quality of life, to the same extent. Families may have other constraints such as finances that may limit the family's ability to maintain the same quality of life. It has been suggested that these issues contribute to the possible exacerbation of anxiety due to decreased quality of life and poor adaptation to this syndrome.

The last psychosocial issue found in the research is the uncertainty that parents expressed about not knowing what to expect in the future with regard to LQTS. This uncertainty has the possibility to affect decision-making because of parents' awareness of the fatal consequences of this syndrome. Parents suggested that this uncertainty guided their treatment decisions. It was also suggested that families were better able to cope and adjust to this syndrome over a period of time. The adjustment that came with time also allowed parents to increase their knowledge about this syndrome, thus contributing to a healthier coping style and approach to this syndrome and (Farnsworth et al., 2006).

The previously mentioned research indicates three main areas of concern for those individuals affected by LQTS. Although this research presents much needed information about concerns within this population, because of the limited sample size and the broad themes there is a need for increased research. Further research with a larger sample size is needed to delineate how these specific areas of concern and other specific factors can contribute to anxiety.

Hendricks et al., (2005) presented initial research on coping and adjustment in relation to LQTS. Researchers recognized the lack of research and the unique aspects of the syndrome that may contribute to varying outcomes in relation to psychosocial aspects of LQTS. Upon initial diagnosis, 50% of parents indicated significant levels of distress. Those parents who were more familiar and had previous family experiences with the syndrome had higher distress levels in comparison with parents without much previous

knowledge or experience (Hendricks et al., 2005). This initial research on LQTS provided impetus for understanding more long-term psychosocial consequences.

Hendricks et al. (2005) completed an additional study eighteen months later to assess these long-term outcomes in parents with adolescents with LQTS. These results suggested that eighteen months after genetic testing was completed that there was still significant distress. This emotional turmoil was increased in parents with lower education, higher anxiety score on research measures, those individuals familiar with the syndrome for a longer period of time, and parents who experienced a loss of a family member from LQTS. It was also suggested that the quality of information received was an important component of anxiety related to LQTS. Many of the parents indicated similar themes of anxiety that contributed to their continued levels of distress (Hendricks et al., 2005)

Seventy-five percent of 36 parents indicated anxiety as a result of feeling as though the syndrome was a constant threat. Parents suggested that they had continuous thoughts that their adolescents could develop life threatening symptoms at any juncture, even with adherence to medical regimens (Hendricks et al., 2005). These expressed concerns are consistent with the uncertainty and fear of death that has been seen in previous research. Even with strict adherence to medical recommendations, there still remains a sense of danger that continues to elicit anxiety in parents and the adolescents. Interestingly, 20% of parents indicated full confidence in medical care; however, over 50% of parents found treatment to be burdensome. This burden speaks to quality of life and the level of adaptation that this syndrome requires to continue a satisfying lifestyle. Individuals affected by LQTS make significant lifestyle modifications to adhere to medical recommendations in order to prevent cardiac events. Another source of anxiety

indicated by parents concerned the future of their children. The anticipation of negative consequences of this syndrome was pervasive. They indicate fears related to their adolescents' careers, relationships, and overall quality of life (Hendricks et al., 2005).

A last major element of anxiety for parents was related to information received about LQTS. Fifty-five percent of parents were unhappy with their ability to access information about the syndrome. Parents also suggested poor support, overall, from the medical community, and sometimes felt as though they provided more knowledge to their doctors than they received (Hendricks et al., 2005). This perceived lack of support within the medical community directly contributes to anxiety because of poor access to information and thus to the parents' ability to understand the overall long-term consequence of this syndrome. The lack of interaction between parents and the medical community promoted uncertainty that appeared to contribute to anxiety in parents. Overall, this research suggests the need for specific education and clear instructions on how to manage LQTS effectively. The results of this study suggest that many healthcare providers lack specific knowledge about this syndrome, which contributed to increased fear and uncertainty in parents (Farnsworth et al., 2006). This perceived lack of information could contribute to anxiety if a family feels overwhelmed and uncertain about treatment.

This specific study speaks to themes similar to those addressed in earlier research, in addition to more specific variables that related directly to anxiety. This research begins to glean more specific variables directly related to symptomology of LQTS. It is important to begin to understand this syndrome as being unique from other chronic illnesses because of the lack of knowledge and because of the fatality of the symptoms. As more specific factors related to this syndrome are identified, there is increased

opportunity to understand how to address anxiety effectively in this population. Similar to the other study, an increased number of subjects are needed to provide data related to the interaction between identified variables and anxiety.

Giuffre, Gupta, Crawford, and Leung (2008) presented additional research about the psychosocial aspects of LQTS. It was suggested that families and individuals affected by this syndrome would experience significant fear and anxiety related to the uncertainty and to the life threatening aspects of the syndrome. This research presented a comparison between a similar life threatening chronic illness (asthma) and LQTS, including whether or not there was increased anxiety due to chronic illness and fear of death. Researchers suggested that those affected by LQTS would present more anxiety and fear concerns than those affected by asthma, because of the differences in severity levels of the illness and differing vulnerabilities (Giuffre, Gupta, Crawford, Leung, 2008).

Results indicate that children with asthma express strong fears related to danger, death, and minor injury. Children with LQTS do not indicate these same fears possibly because of the difference in physical symptomology between asthma and LQTS. Children with asthma have recurrent episodes of difficulty in breathing and other related physical sensations. In comparison, children with LQTS have limited symptomology that present different aspects of severity and uncertainty related to their chronic illness (Giuffre et al., 2008). The discrepancy between chronic illnesses indicates the increased need for an understanding of LQTS. Although past research has suggested an overall generalization of the psychosocial aspects of chronic illness, this research indicates that differing symptomology and courses may lead to different exacerbating factors and consequences.

Giuffre et al. also indicated that children with LQTS may have a higher need for social desirability and that they fear criticism and failure (Giuffre et al., 2008). These fears may be heightened from the lack of understanding of the syndrome within the medical and overall community. In addition, the differing symptomology that is often not understood by others and not recurrent in comparison with asthma may exacerbate these fears. It was also suggested that children with LQTS display high levels of internalizing and externalizing behaviors that suggest overall ineffective coping mechanisms. These adolescents may be unable to express feelings appropriately; rather, they may isolate or act out in different environments. This presents initial suggestions of how one would approach treatment with these adolescents, but still lacks significant substance that is well understood in relation to other chronic illnesses (Giuffre et al., 2008).

Last, data indicate multiple parental concerns. Their anxiety was elevated in relation to clinical judgment, understanding this syndrome, and compliance. Interestingly, parents had greater anxiety after initial diagnosis and after dealing with the syndrome for a greater extent of time (Giuffre et al., 2008). This presents an interesting time line of anxiety for parents. Prior research of chronic illness has suggested initial anxiety that dissipates with time and knowledge. The renewal of anxiety after a period of time suggests the realization of a lack of knowledge and increased uncertainty about how to live and cope with this syndrome.

The development of anxiety can be conceptualized in many ways according to varying theories. Each theory speculates variables that contribute to the development of anxiety. These variables lead to emotional and behavioral reactions in an effort to handle anxiety. Clearly, there has been significant research suggesting the presence of anxiety in

chronic illness populations. Anxiety related to LQTS will be described through different theoretical perspectives to provide a possible conceptualization of anxiety within this population.

Introduction to stress and adaptation.

The concept of stress and adaptation has been refined over time with increased understanding. The conceptualization of stress dates back to the 17th century in terms of physical sciences and the ratio between internal and external forces exerting pressures on an object. In the 19th century, Hans Selye (1936) developed the general adaptation syndrome, which was one of the first organized concepts to explain reactions and processes caused by environmental demands that induce stress reactions. Harold Wolff continued to elaborate upon the idea of stress and emphasized it as a more dynamic concept that started to elucidate ideas of adaptation and coping aspects (Lazarus & Folkman, 1984). Developments such as these initial concepts played a significant role in the development of understanding the concept of stress.

Sigmund Freud (1944) suggested anxiety or stress in the role of psychopathology. Prior to this theory, the broad understanding of anxiety had been utilized to explain all psychopathology. Freud postulated specific mechanisms to suggest the role of anxiety in explaining certain symptomology. Freud's theory suggested that anxiety was a result of an internal drive conflict that created a perception of threat or danger, led to poor coping and thus elicited defense mechanisms (Lazarus & Folkman, 1984). In a sharp contrast to psychodynamic theory, behavioral theorists suggested a more mechanistic concept of anxiety. The behaviorist viewpoint suggested that anxiety was a conditioned response to specific environmental stimuli (Forgays, Sosnowski, & Wrzesniewski, 1992).

Other theories began to incorporate more internal drives that elicit stress responses in an effort to understand the complexities of the human experience of anxiety more fully. These approaches to anxiety described certain personalities as being more susceptible to anxiety. The trait approach to anxiety hypothesized that certain individuals were inherently susceptible to the development of anxiety responses in provoking situations. These types of theories emerged from overarching theories that suggested the commonality of all humans towards the development of anxiety. These personality driven theories suggested a more individualized, internal framework; however, they still lacked an understanding of variation among stressors and anxiety responses. Rather, there was suggestion that at birth an individual either would experience anxiety or not experience anxiety when exposed to similar types of stressors (Forgays, et al., 1992).

These viewpoints lacked an understanding of variables related to specific individuals and cognitive components of anxiety. In contrast, the interactionist model suggests the influence of the brain and mind, or cognition on emotions. The incorporation of cognition into models of stress was a novel approach. However, it was still limited because there was no interaction among the three variables; rather, cognition and the brain indirectly influenced each other through emotion (Aldwin, 2007).

Through previous theoretical underpinnings, theorists further adapted models of emotions to explain the human experience. Although initial theorists attempted to explain the cause of stress, later theorists took an alternate approach. In the 1960s, various world events such as the Vietnam and Korean wars increased awareness that stress is an inevitable factor in all humans' lives. Theorists utilized this knowledge of trigger events that cause stress and shifted their focus towards adaptation and coping. Researchers began to question the reasons why some individuals, when confronted with

similar stressors, were able to handle the situation adaptively, but others exhibited more dysfunctional responses, including anxiety (Lazarus & Folkman, 1984).

Cognitive models.

Mason (1975) eliminated much of the ambiguity obscuring the concept of stress. He suggested three variables in which the concept of stress could be utilized in order to understand situational and individual reactions to trigger events. First, he suggested that stress could refer to an internal state. These internal strains could be physiological or emotional reactions. Often these emotional reactions refer to anxiety, anger, guilt, or depression that is caused by significant life stressors. It is suggested that individuals affected by chronic illnesses may, during the course of the illness, experience these emotional reactions, specifically anxiety (Aldwin, 2007). As an individual continues to have experiences related to a chronic illness, there may be an increase or decrease in anxiety or physical reactions to hospitalization or other illness related events.

The next variable that in early theories would have been considered a separate entity is the external events or stressors. There are many important considerations related to stressors: the type of stressor, the duration, and the circumstances at the onset (Aldwin, 2007). Chronic illness, in regard to these variables, brings forth interesting dynamics for consideration. Previous thought had been that a general conceptualization and model of chronic illness was enough to understand various adaptation and coping strategies of individuals. One might consider parents with older adolescents; they experience a fatal event, suddenly, without knowledge of their child having LQTS, and then discover that their other children also have LQTS and these children have to receive life-altering treatments. The rapidity of onset without previous knowledge of a relatively unknown syndrome that is fatal has been their only experience with this syndrome. This

suggests a response possibly different from another family, whose child is diagnosed with a well-researched and maintained chronic illness. There should also be consideration of the interactions between the internal and external responses. Previous experience and knowledge with stressors is going to affect one's overall reaction and ability to handle certain situations, in addition to feelings of competence. Lack of competence and perceived inability to handle illness related external experiences are going to affect internal and external physical reactions (Aldwin, 2007).

The last variable defined by Mason is interactional and considers both the person and environment. There are two components of this variable: cognitive appraisals and intensity of the stressors. Stress is experienced differently, based on cognitive appraisals of the situation. Different appraisals can include harm or loss, threat, challenge, controllability, or neutrality. Lazarus and Folkman have suggested that negative emotional or physiological reactions occur when there is a perception of threat, harm, or loss. Lazarus and Folkman provided original ideas that suggested stress etiology should account for internal and external events. Also, an individual's perception and appraisal of events that provoke feelings either of competence or of incompetence in dealing with the stressors, will lead to anxiety or will buffers anxiety and alternate behavioral ramifications (Aldwin, 2007). The incorporation of cognitive appraisals and varying levels of stressors increased the understanding of stress, and how the relationship between an individual and his or her environment can lead to negative psychological consequences, based on cognitive appraisals of the situation (Lazarus & Folkman, 1984).

Cognitive models of coping incorporate the concepts of stress, coping, and adaptation and describe the interactional nature of these three variables. All cognitive models of coping operate on four assumptions. The first assumption is that individuals

consciously choose how to cope, based on their appraisal of the situation. These cognitive appraisals relate to Mason's idea of transactions between the environment and individuals. Based on whether or not a person appraises a situation as harm, loss, threat, or challenge affects his or her overall coping response. Those situations that are deemed neutral will cause no necessity for coping because it is not needed. If a stressor is viewed as a threat or loss, the individual may choose an approach that oriented toward problem solving or may experience an adverse psychological reaction such as anxiety. This assumption rests on the fact that humans are thinking beings that make conscious decisions on how to cope, based on previous experiences. In addition, individuals account for a situation being changeable or unchangeable to determine the type of coping that would be most effective (Aldwin, 2007). Individuals affected by LQTS may display a developmental process related to coping. It is suggested that initial experiences with this syndrome may evoke feelings of helplessness or lack of control that can lead to anxiety. As a family becomes more aware of the process associated with this syndrome, their overall coping may become more adaptive, or feelings of helplessness and lack of control may remain because of an inability to utilize appropriate adaptive resources.

The second component of the cognitive model is that coping is situation-specific. Unlike previous theories that suggest a uniform approach to coping in all situations, cognitive theories suggest that coping is a situational variable. Whereas the previous assumption accounted for individual differences in coping, this assumption represents varying environmental contingences that may affect coping and adaptation (Aldwin, 2007). For instance, individuals affected by LQTS may cope effectively in academic environments because of the levels of support received by teachers and other staff members. Conversely, individuals with LQTS may find difficulty coping in medical

environments because of the lack of support and knowledge provided by medical professionals. However, other individuals affected by LQTS, may have different situational variables in these environments, which affect their coping abilities.

The next component of the cognitive model involves problem and emotion focused coping strategies. There are no assumptions as to which strategy is better utilized in particular situations. It has been suggested that problem focused strategies often lead to better overall emotional control (Aldwin, 2007). An individual with LQTS might display emotion focused coping strategies when they receive limited information about the syndrome; they may feel a sense of helplessness that leads to anxiety. A problem focused strategy would be to take an active approach to seeking out information and resources related to the syndrome.

The fourth assumption of the cognitive model is the lack of a hierarchical structure of coping strategies. Individuals use an empiricist approach to select the most effective coping mechanisms, based on situational variables. It has been suggested that certain strategies such as denial work very well in some situations but not in others. Similar to other assumptions, the interaction of the individual and environment are considered before assuming a certain strategy of coping that will lead to the overall best adaptation (Aldwin, 2007).

Coping is broadly described as a reaction to any stressor in which the perceived demands seems to outweigh the perceived resources. With time, this definition was expanded, because it lacked several variables that also contribute to an individual's ability to cope. Coping can be a maladaptive or adaptive response to certain triggers. In order to truly conceptualize an individual's response, an understanding of all contributing factors and interactions must be taken into consideration. A coping response is based on

individual and illness-specific factors that contribute to a person's perceived ability to handle aspects of the illness (Zehnder et al., 2006).

Coping has been further broken down into three variables that account for variance among coping responses. The first variable is proactive responses to stress (Compas, 1987). With increased knowledge, individuals engage in active coping responses (Lazarus & Folkman, 1984). This concept of actively responding to stress situations indicates effort and lack of pure instinctual drives in relation to coping processes. These efforts to cope with stressors account for successful and unsuccessful attempts in negotiating stressful situations. With these attempts, an individual may change his or her cognitive and behavioral patterns to meet the demands of the situation, based on perceived resources, to handle the stressor. These modifications in coping processes suggest constant appraisals that are influenced by individual perceptions (Compas, 1987).

The function of coping should also be considered when defining the construct of coping. There are two functions of coping that can lead to adapting to a stressor. Acting on a stressor indicates strategies, such as problem solving or modification of the relationship between the individual and stressor. The second function of coping is emotion regulation through strategies of avoidance, cognitive reframing, or selective attention (Compas, 1987).

The next variable, resources, style, and specific coping efforts, are also important to the discussion of coping. This variable accounts for varying levels of resources available to specific individuals. It includes internal resources, such as problem solving and interpersonal skills, and the influence of self-image on aspects of control, assertiveness, and decision-making. Resources also include external means such as

social support or medical assistance. Resources allow for increased adaptation to stressors. When these resources are limited they can lead to poor outcomes on psychological and environmental levels.

Coping styles indicate a particular manner in which a person handles stressors; these are influenced by values, beliefs, and goals. These styles may also be influenced by cognitive appraisals of situations, such as the belief of controllability in certain environments. Last, specific coping efforts are cognitive and behavioral styles that have been used in past, similar situations or for an ongoing stressor. These efforts may vary, based upon present circumstances and past experiences (Compas, 1987).

Adaptation is a comprehensive term that includes concepts such as mastery, defenses, and coping strategies. The concept of adaptation conceptualizes how individuals successfully cope not only with everyday life stressors, but also with more chronic and long lasting events (Aldwin, 2007). A basic definition of adaptation in relation to biological systems provides a very simplistic point of reference as a starting point in understanding more complex adaptation processes of humans. Biological principles suggest that successful adaptation of an animal or plant is the capacity of the organism to survive and flourish. In order for the organism to adapt successfully, it must have the proper environment to provide food, nutrients, and other basic needs. As an organism becomes more complex, basic, life sustaining elements are not enough to adapt successfully to more complex environments. Adaptation becomes a more specifically interactive process that includes physical, psychological, and social aspects. These aspects must be attended to in order to understand adaptation fully, including how one adapts successfully to varying stressors and environments (Lazarus & Folkman, 1984). It has been suggested that individuals with anxiety disorders have adapted poorly to

stressors. An understanding of unique illness aspects, which contribute to the experience of anxiety, offer areas for intervention to help persons affected by LQTS to improve their coping skills and adapt to this condition.

Transactional stress and coping model.

Lazarus and Folkman presented one of the first theoretical frameworks used to understand varying coping responses of individuals presented with similar trigger events. Lazarus and Eriksen (1952) provided initial evidence for this shift in focus towards individual coping responses. This framework was first developed in response to varying stress responses exhibited by individuals who experienced failure. This research suggested that under similar stressful situations, individuals have alternating levels of stress that could be predicted only through internal processes (Lazarus & Folkman, 1984). This initial study highlighted the movement away from the interactionist model towards the transactionist model. This model incorporates the same three variables as the interactionist model; however, it highlights the interaction and influence that all three variables have on each other in the process (Aldwin, 2007).

There are two major elements to a transactionist point of view. The first element is that all three variables influence each other to the same degree. This model elucidates how the brain can influence cognition or how things that influence cognition such as society, interactions, or culture can also directly influence biological mechanisms. The second component accounts for previous experience in current coping mechanisms. Researchers hold varying opinions about how past experience can interact with current coping processes. Researchers agree on the idea that that a developmental process occurs both in positive and negative stressful interactions, affecting future adaptation and coping efforts (Aldwin, 2007).

The development of this theory led to the recognition of a lack of understanding about the concept of stress and its consequences. Whereas many individuals suggested that stress is a generic term that has little value in the conceptualization of human processes and behaviors, Lazarus suggested a more sophisticated viewpoint. He suggested that stress could serve as an organizational concept in the understanding of multiple variables and processes that occur within individual's lives. His developmental theory was able to provide a framework that would allow for the examination of possible triggers, behaviors, and consequences of stress related phenomena. The recognition of varying triggers that elicit stress responses was a novel concept and provided impetus for the conceptualization of individual factors of stress, providing reasons why some individuals cope with stress adaptively, yet others develop anxiety and other psychosocial implications (Lazarus & Folkman, 1984).

Lazarus and Folkman (1984) utilized the transactional framework to explain the coping process that evolves when an individual is confronted with a stressful situation. The variables included in this coping model are stress, personality, and coping. The same assumptions hold true as in previous transactionist models; they mutually affect one another and a developmental process occurs. However, the variables allow for the continual evolution towards an understanding of the coping process. For instance, stress can be viewed as an environmental demand that influences a person's personality, or someone's personality can affect how one handles an environmental demand. Examination of this from a developmental perspective suggests how previous coping strategies influence the degree to which an individual perceives he or she has control (or lack control) of a situation (Aldwin, 2007).

Application of this model to LQTS suggests several possible interactions and outcomes. Stress or the environmental variables may be related to experiences in academic, family, or medical settings that may, in turn, be influenced by lack of previous experience in this setting, relative to chronic illness, or it may be influenced by previous experience that suggests a complete lack of control over the syndrome or decisions. Or an individual may have personality characteristics that prevent effective decision making or problem solving that, in turn, influence not only the stressor, but also how one attempts to cope with a situation. The perceived inability to cope with a situation effectively may be exacerbated by stressors, personality, previous experiences, or an interaction of any other variables. The selected coping strategy produces either an adaptive or a maladaptive mechanism of action. The goal is to understand, further, specific variables that contribute to anxiety within this population. The multiple interactions among variables can become extensive and elaborate in nature. Identification of common stressors or environmental variables experienced by individuals affected by LQTS would be helpful in conceptualizing the types of anxiety symptoms they experience, so that coping and adjustment can be studied systematically in the future.

A major component of the transactional model is the role of cognitive appraisals on the process of handling stressors and resulting consequences such as anxiety. The role of cognitive appraisals was developed and has led to an understanding of varying responses to similar external conditions. The concept of appraisal recognizes that situations are based on judgment, selection and choices based primarily on past experiences. These appraisal processes recognize that cognitive processes are a result of a unique combination of individual factors interacting with the event and overall outcome (Lazarus & Folkman, 1984).

There are two major types of appraisal processes: primary and secondary. Primary appraisals are the process of perceiving a threat in relation to oneself. Through this process, an individual can appraise a situation in three ways. An irrelevant appraisal suggests that the situation has no implications for his or her personal well-being. The second type of primary appraisal is benign-positive. This type of appraisal is perceived as positive and promotes the well-being of the client (Lazarus & Folkman, 1984). For example, it could be suggested that families affected by LQTS may perceive the implantation of an ICD as benign-positive because of the sense of security it provides to the family in relation to life ending symptomology.

The next type of primary appraisal is stress appraisals. Stress appraisals include three types: harm/loss, threat, and challenge. Harm/loss appraisals suggest that an individual perceives a harm/loss in relation to the situation. An individual may have lost certain functional abilities due to an illness, or lost a significant other to a traumatic event or disease. Threat appraisals are anticipatory reactions to perceived harm or loss. In comparison with harm/loss appraisals, threat appraisal allows an individual to engage in coping strategies prior to the event. However, the perception of a threat often suggests that an individual feels unable to cope adaptively with the situation, or that the threat may be real (Lazarus & Folkman, 1984). Hendricks et al. (2005) suggest that LQTS is often appraised as a threat. These appraisals have led to overwhelming anxiety in 75% of the 36 parents of adolescents with LQTS. This appraisal process leads to coping efforts, which include continual worry about consequences, and behavioral strategies, such as constant reassurance from adolescents. This research implies the need for continued education about this syndrome and about effective treatments (Hendricks et al., 2005). An individual affected by LQTS may appraise the situation as threatening because of

limited knowledge and about resources related to the syndrome. This perception may lead to anxiety because of the inability to use resources effectively in order to engage readily in proactive strategies. Yet the literature does not offer a comprehensive understanding of those LQTS related situations, which might be perceived as threatening.

The last type of stress appraisal is challenge. Similar to threat appraisal, challenge appraisals provide the opportunity for coping efforts. Unlike threat appraisals, the focus moves away from an inability to cope, to the perception of ability to overcome the precipitant effectively and provide opportunity for learning and growth. Those individuals who experience challenge versus threat appraisals may experience varying emotional states as well. Challenge may lead to more positive emotions, including excitement and eagerness, whereas those perceiving a stimulus as a threat experience fear and anxiety (Lazarus & Folkman, 1984).

Secondary appraisals are the process of developing potential responses to stimuli, such as treatment decision in LQTS. These are based on primary appraisal, which guides decision making on how to handle and cope with the situation effectively. Based on these perceptions, secondary appraisals account for coping options and the probability that these options will lead to a successful outcome. An individual also has to feel that he or she is competent in utilizing these strategies effectively and that the overall outcome will be beneficial. Thus, understanding LQTS-specific anxiety provoking triggers will help professionals better educate and prepare persons affected by LQTS for better coping. These beliefs about outcome and efficacy expectations are essential to conceptualize how an individual will enact coping efforts (Lazarus & Folkman, 1984). An individual with LQTS may perceive the efficacy of his or her actions to be low, based on past outcomes within the medical community, and thus choose a more passive form of coping than those

who perceive past efforts as efficacious. An individual's competence to handle and cope effectively is essential for positive outcomes.

Secondary appraisals also can lead to emotional consequences for an individual. If an individual feels competent in handling a perceived challenge, he or she may feel inspired or motivated. In comparison, an individual that has the perception that efforts to overcome the problem are limited or will be ineffective may endure anxiety or other negative emotional implications (Lazarus & Folkman, 1984). These perceptions of limitations and effectiveness are pervasive within the chronic illness population. This suggests the role of appraisals in relation to anxiety through feeling a lack of control and a vulnerability to symptoms of this syndrome (Giuffre et al., 2008). Thus, identifying anxiety-provoking triggers and helping persons prepare for them may increase their sense of competence.

The third component of the transactional model is the integration of coping, based on the outcomes of the appraisal processes. Coping is the execution of the response, based on the perception of the stimuli and overall belief about the effectiveness of certain responses (Carver, Scheier, & Weintraub, 1989). Coping represents cognitive and behavioral efforts that are intended to mediate external and internal stressors that are perceived to exceed or burden an individual's personal resources. These coping efforts represent a strategy that leads to an expected outcome for an individual. These outcomes may be mediated by appraisal processes and through selection of specific types of coping strategies (Lazarus & Folkman, 1984). These efforts to cope with stressors are not always successful in mediating the situation and can lead to increased emotional consequences, such as anxiety (Compas, 1987).

The transactional model discusses two forms of coping: emotion-focused and problem focused. Emotion focused coping is elicited in an attempt to manage emotions created by a precipitant. This type of coping is often prevalent when an individual feels that not many actions can be taken to alleviate the precipitant, but that he or she must cope with the emotions in some fashion (Carver et al., 1989). Carver, Scheier, and Weintraub (1989) found that individuals engaged in emotion-focused coping in uncontrollable situations that were of significant importance to the individual (Carver et al., 1989). Different emotion focused strategies may take the form of avoidance, minimizing, unfair comparisons, or selective attention to certain aspects (Lazarus & Folkman, 1984). Emotion-focused strategies may focus too much on emotions, and prevent adjustment to a stressor and action (Carver et al., 1989). Aldwin and Revenson (1987) utilized the transactional model to understand the relationship between coping and mental health consequences. It was suggested that emotion-focused coping types were avoidance, blame, minimization, and finding meaning through the experience. Maladaptive coping was more often viewed in individuals with previous emotional distress; these individuals have more current, significant stressors that tended to elicit maladaptive coping strategies and further emotional distress. These coping strategies were more often emotion focused; in particular, avoidance and blame tended to elicit more emotional distress (Aldwin & Revenson, 1987).

Individuals affected by LQTS may focus on emotional consequences of the syndrome when they perceive themselves as having little control over the diagnosis, condition, or management of LQTS. Individuals may also disengage through avoidance or distraction in regard to aspects of the syndrome. These avoidance strategies serve to alleviate anxiety in the short term; however, they do not result in strategies that will

alleviate anxiety in the long term. Denial may also be utilized with emotion-focused coping. At times, this minimization may serve to alleviate anxiety. It can also lead to more difficulties such as medical complications or poor overall functioning. Individuals may also alleviate distress related to the syndrome by acceptance of the diagnosis and consequences. This acceptance may serve as beneficial, initially; however, individuals may not learn about alternative approaches to treatment or may not learn how to handle the syndrome effectively so that anxiety could be alleviated in the long term (Carver et al., 1989). Identifying common, anxiety-generating experiences in the LQTS population will allow professionals to increase knowledge and resources surrounding these issues in order to facilitate an adaptive appraisal of one's ability to respond to these stressors. Individuals may feel more competent in their abilities to alter the situation through a more problem-oriented approach, based on primary and secondary appraisals.

Problem-focused coping is action oriented. This type of coping is oriented towards problem solving and modifying sources of stress. Individuals that engage in this type of coping feel as though actions can be taken to alter the source of stress. These individuals may appraise the precipitant more as a challenge than as harm/loss or a threat (Carver et al., 1989). Individuals that engage in this type of coping often define the problem, generate solutions, and implement a decision based on the pros and cons of varying solutions (Lazarus & Folkman, 1984). These individuals also may engage in planning activities that allow them to develop clear and effective strategies on how to handle a stressor (Carver et al., 1989). The research of Sales, Fivush, and Teague (2008) displays the beneficial aspects of parental coping styles that result in decreased anxiety in them and in their asthmatic adolescents. Overall, mothers who engaged in more active coping strategies in contrast to avoidance coping strategies displayed lower levels of

anxiety, and their adolescents indicated lower anxiety and higher quality of life. These families also had fewer illness restrictions and their healthier, overall emotional functioning exceeded that of more avoidant families (Sales, Fivush, & Teague, 2008).

Within the LQTS population, second opinions about medical recommendations may be sought, or strategies designed to alleviate worry when a child is not within the parents' care may be planned accordingly. Individuals utilizing this type of coping may also delay action until a more appropriate and effective time. This strategy is not passive; rather it is considered active, because the time delay allows for enactment of more effective problem solving approaches (Carver et al., 1989). Individuals affected by LQTS may display this type of behavior when choosing to get alternative medical advice before performing invasive procedures. Support seeking is also considered a problem-oriented approach, whether for seeking information or for emotional support. The utilization of social support for emotional disclosure may lead into emotion focused coping if it does not allow an individual to actively engage in other problem oriented strategies, and, rather, remains focused on distressing emotions (Carver et al., 1989). Identifying the experiences of anxiety in the LQTS population will help patients identify, in advance, times for which they may need to prepare in order to rally additional support or to prepare fully to engage in problem-solving activities.

Research suggests that adolescents with chronic illnesses are at increased risk for anxiety and other adjustment difficulties when they engage in emotion-focused strategies related to their illness rather than engaging in problem-oriented strategies (Sender, Prchal, Vollrath, & Landolt, 2006). Meijer et al. (2002) suggest that chronically ill adolescents that engage in active problem solving strategies had less anxiety than those adolescents engaging in less behavioral oriented strategies. This research also has

implications for the LQTS population in relation to the identifiable coping strategies that work well within other chronic illness populations and applying these strategies to LQTS treatment (Meijer, Sinnema, Bijstra, Mellenbergh, & Wolters, 2002). It is suggested that individuals working with this population would initially need to identify common factors that elicit stress responses and anxiety in this population.

Zehnder, Prchal, Vollrath, and Landolt (2006) suggested that within a chronically ill adolescent population consideration of various factors should be addressed before assuming that one type of coping mechanism is more adaptive than another. This research identified the fact that emotion focused strategies, such as distraction and avoidance, are effective for decreased psychosocial consequences in the short term and in uncontrollable situations, as previously suggested by the transactional model (Zehnder et al., 2006). This research has substantial implications for those affected by LQTS in relation to the level of control and coping mechanism selected to lead to successful adaptation to this syndrome. Individuals affected by LQTS experience overwhelming emotions when trying to adjust to LQTS. It is suggested that a healthy moderation between emotional and problem focused strategies may be most adaptive when an individual has support systems, but continues to follow medical recommendations and engages in adaptive lifestyle modifications.

Often an individual selects coping methods based on appraisals of the resources available to him or her and in consideration of restraints on these resources, based on the type of precipitant (Lazarus & Folkman, 1984). In relation to coping, these resources are especially important to conceptualize in relation to LQTS, because the significant precipitants that can tax individuals and limit their resources for engaging in coping mechanisms evoke feelings of threat and vulnerability, contributing to anxiety.

Experiences of adolescents coping with chronic illness can be conceptualized somewhat differently from adults affected by LQTS (Compas, 1987). It is important to conceptualize adolescents' experiences in terms of their family unit, in conjunction with their own developmental process and situational factors. A main determinant of coping is the developmental level of the adolescent. The selection of strategies is based on the resources available, in addition to cognitive, physical, and emotional abilities that are highly dependent on the developmental processes. Adolescent developmental levels also suggest age appropriate coping that lead to more advanced strategies through utilizing more resources and emotional control (Hutchinson et al., 2006). Emotion focused and cognitive strategies increase with age and increased cognitive functioning (Zehnder et al., 2006).

The interaction of resources available to an individual represents the vulnerability to the development of anxiety in those affected by LQTS. The ability to cope adaptively is based on numerous factors that have to be implemented effectively when confronted with a chronic stressor (LQTS). It has been suggested that the inability to utilize these skills effectively contributes to anxiety because of a lack of resources or because of inability to implement coping skills. The consideration of various resources and the totality of the dynamics of the coping process can lead to the selection of certain strategies. As suggested previously, conceptualizing the entire picture can lead to the view of controllability or uncontrollability of illness related stressors, which directly contributes to uncertainty and anxiety about future consequences of this syndrome (Weisz, McCabe, & Dennig, 1994). The recognition of interaction of factors that lead to successful adaptation to LQTS is essential. These interactions occur between illness experiences, individual and family factors, and processing of stressors. The role of

appraisal and beliefs on overall coping methods leads to an understanding of the development of anxiety and other consequences of poor coping skills (Thompson & Gustafson, 1996).

Improving the understanding of chronic illness and successful coping and adaptation has led to the development of interventions. These interventions have recognized the need for the development of specific coping skills and interventions when confronted with a chronic illness. Bandura (1986) provided significant rationale for coping skills training that suggests that practicing and rehearsing new behaviors can lead to improvements in coping. These improvements also influence self-efficacy and confidence that decrease the likelihood of anxiety and other emotional consequences (Davidson et al., 1997). Hampel et al. (2005) suggest that chronically ill adolescents who had underdeveloped coping skills were at greater risk for psychological difficulties (Hampel et al., 2005).

Coping skills training has been implemented in multiple chronically ill populations. Adolescents with diabetes endure multiple stressors from adapting to the illness through the developmental process and interacting with peers and family in appropriate ways. Coping skills training that has utilized cognitive skills and assertiveness training has been successful in leading to adaptation to their chronic illness (Davidson et al., 1997). Grey et al. (1998) implemented coping skills training for adolescents with diabetes. Adolescents, who appeared to benefit from learning coping skills, appeared to have better medical management of their diabetes. Poor psychosocial adjustment led to poor medical management in regard to self-monitoring, diet, and daily injections. This intervention sought to influence adolescents' belief systems about their overall efficacy in managing illness-related stressors. Results suggest an increased

psychosocial well-being that resulted in adolescents feeling more efficacious and competent in handling their chronic illness. Specific strategies allowed adolescents to perform successfully in multiple areas of their lives. Mastery of these skills influenced their beliefs that they could control aspects of their chronic illness through effective, focused coping skills. There was also a noticeable change related to these adolescents' social and emotional adjustments after the development of coping skills (Grey et al., 1998).

The understanding of ineffective coping and adaptation to chronic illness is a critical theoretical element in conceptualizing how anxiety develops into anxiety disorders. Specifically, the role of appraisal processes and specific coping strategies allows for the conceptualization of how anxiety disorders can develop within the LQTS population. Other factors may subsequently cause anxiety-related responses and these are addressed in the following section.

Overview of anxiety.

Anxiety can be conceptualized as a protective biological mechanism by mentally and physically alerting someone to danger (Emilien, Durlach, Lepola, & Dinan, 2002). It has been suggested that a moderate amount of anxiety enables individuals to direct their attention to salient stimuli and enact appropriate coping behaviors. Maladaptive anxiety is associated with highly negative effects. This level of anxiety has associated cognitive and behavioral manifestations. Cognitive aspects of anxiety are constant hypervigilance, biased awareness of possible threats, and poor concentration on everyday tasks. Maladaptive anxiety is manifested behaviorally through increased fear of specific behaviors, increased startle response, and constant readiness to respond to negative events (Emilien et al., 2002).

These behavioral and cognitive manifestations of anxiety also prohibit someone from engaging in adaptive coping techniques. Dembo, Leviton, and Wright (1956) suggested a successful coping framework that is useful in conceptualizing how anxiety becomes maladaptive from a chronic illness standpoint. This framework includes the following characteristics: an understanding of one's capabilities, the ability to be active within one's life, recognition of accomplishments, the ability to handle negative situations, decreased limits through environmental modification, and participation in pleasant activities (Livneh & Antonak, 1997). It becomes clear that maladaptive anxiety decreases an individual's ability to function within his or her daily life (Emilien et al., 2002). It is suggested that a further understanding of the anxiety triggers for people with LQTS is needed to understand specific cognitive and behavioral manifestations of anxiety in this population.

In order to understand more fully how anxiety can affect these areas of coping that are related to chronic disease, it is helpful to conceptualize anxiety based on a categorical perspective. The symptoms of anxiety are listed under two categories: subjective and objective. The subjective symptoms of anxiety are psychological experiences that include: worrying, terror, mental acts such as thoughts of dying, fear, and depersonalization. The objective symptoms of anxiety are often referred to as somatic and include pain, dizziness, palpitations, hyperventilation, hot flushes, dry mouth, headaches, and restlessness.

The separation of these symptoms can also be based on cognitive, affective, physiological, and behavioral changes. Cognitive aspects are related to sensory-perceptual symptoms including: unreality, hypervigilance, thinking problems, fear related beliefs and thoughts, and depersonalization. Affective changes of anxiety

include: intense feelings of guilt, shame, anger, apprehension, and fear. Physiological changes related to anxiety result from hyperarousal to prepare for fight or flight. At the behavioral level, individuals with maladaptive anxiety may exhibit decreased behavioral responses that are related to safety, avoidance, or helplessness (Emilien et al., 2002).

This broad understanding of anxiety can be further broken down, based on the American Psychiatric Association Diagnostic and Statistical Manual of Mental Disorders-Fourth Edition-Text Revision (DSM-IV-TR).

Panic disorder with and without agoraphobia.

Panic disorder occurs at times when an individual experiences intense pain or discomfort with four or more of the following symptoms: palpitations or accelerated heart rate, sweating, trembling or shaking, shortness of breath, feelings of choking, chest discomfort, nausea or abdominal distress, dizziness or lightheadedness, derealization, fear of losing control, dying, or going crazy, numbness, and chills and hot flashes. During the course of these panic attacks, an individual may also develop agoraphobia; i.e., they avoid situations where escape is difficult or help may not be available during a panic attack. The individual develops agoraphobia because of anxiety about having additional panic attacks (American Psychiatric Association, 2000).

Specific and social phobia.

Specific phobia is diagnosed according to the DSM-IV-TR (2000) when there is excessive and persistent fear related to a specific object or situations. The exposure to the specific stimulus creates overwhelming anxiety that is specifically bound to that object or situation. An individual who recognizes that his or her fear is excessive, however, either avoids the stimuli or endures exposure with extreme anxiety (American Psychiatric Association, 2000).

Social phobia is marked by an excessive or persistent fear of social situations at times when there is exposure to unfamiliar people. The individual fears judgment by others and has fears of acting in humiliating ways in these situations. Similar to specific phobias, these situations are either avoided or are endured with extreme discomfort (American Psychiatric Association, 2000).

Obsessive-compulsive disorder.

According to the DSM-IV-TR (2000), obsessive-compulsive disorder is marked either by obsessions or compulsions. Obsessions are recurrent and persistent impulses, images, or thoughts that are viewed as intrusive and inappropriate, thus causing increased anxiety. Obsessions are not about real life worries and individuals may recognize that they are products of their minds. Actions may be taken to try to rid themselves of these obsessions through other thoughts or actions. Compulsions are mental acts or repetitive behaviors that are in response to an obsession. These acts are completed to decrease distress or prevent a negative event from occurring. Compulsions are not realistically connected to the event that an individual is trying to avoid. Obsessions and compulsions cause a significant amount of distress and interfere with normal functioning (American Psychiatric Association, 2000).

Posttraumatic stress disorder and acute stress disorder.

Posttraumatic Stress Disorder (PTSD) occurs after witnessing or experiencing a traumatic event that may have involved feelings of fear, helplessness, or horror. The individual re-experiences this traumatic event in one of the following ways: recurring and distressing recollections through images, thoughts, or perceptions, distressing dreams, acting or feeling as though the event is happening again, intense distress to trauma related cues, or physiological activity related to trauma related cues. An

individual also experiences avoidance and suffers a numbing effect through three of the following ways: avoids thoughts, feelings, and talking about trauma related aspects, avoids activities, places, or people related to trauma, is unable to recall trauma aspects, has decreased interest in activities, detaches from others, has decreased affect, or limited future orientation. An individual must also experience increased arousal in two of the following ways: problems with sleep, irritability or anger, poor concentration, hypervigilance, or an increased startle response. An individual is diagnosed with this after experiencing symptoms for at least one month (American Psychiatric Association, 2000).

Acute stress disorder is also diagnosed after witnessing a traumatic event. An individual must experience three of the following symptoms: decreased emotional responsiveness, numbing or detachment, decreased awareness of environment, depersonalization, and the inability to recall trauma aspects. The trauma must also be experienced in one of the following ways: flashback, images, dreams, thoughts, feelings of reliving trauma, or distress related to trauma cue. The individual also must show significant avoidance of trauma stimuli and symptoms of increased arousal and anxiety. An individual that experiences these symptoms within the first four weeks of the trauma is diagnosed with acute stress disorder (American Psychiatric Association, 2000).

Generalized anxiety disorder.

According to the DSM-IV-TR (2000), generalized anxiety disorder (GAD) is diagnosed after six months of persistent anxiety and worry about numerous events. The individual has difficulty controlling his or her worry. Diagnosis of GAD must also include three of the following symptoms: fatigue, restlessness, poor concentration, muscle tension, sleep problems, or irritability (American Psychiatric Association, 2000).

Conclusion.

From a cognitive perspective, all of these varying symptoms are manifested from selective attention, interpretation, and memory of stimuli within environments that are considered threatening. An individual with maladaptive anxiety makes faulty interpretations about dangerous stimuli within his or her environment. These interpretations are based on the selective processing of certain stimuli and underestimating personal coping strategies and resources. This represents the main difference between adaptive and maladaptive anxiety. An individual with adaptive anxiety is responding to objective threats within the environment rather than to an overestimated perception of danger that is not based on accurate representations of internal and external environments.

In summary, chronic illness literature suggests that persons with serious or life threatening conditions experience heightened anxiety surrounding disease-specific events and circumstances. Appraisal of circumstances and perceptions of competence and self-efficacy to handle such demands affects how individuals cope with their chronic or life threatening conditions. The focus of the current study is to identify common triggers of LQTS-related anxiety and the types of anxiety symptoms experienced by people with LQTS; its further focus is to stimulate additional research on coping with LQTS and to offer areas for medical and mental health professionals to better prepare persons with LQTS with information, predictable challenges on coping with LQTS, and possible support interventions.

Research question.

It is hypothesized that this research will identify specific factors or illness experiences within the LQTS population that lead to anxiety. These factors will be ascertained by focusing on the following research question:

What are the experiences of anxiety in individuals affected by LQTS?

Chapter 3

Methods

Overview.

The current study focused on individuals who have been affected by LQTS and is part of a larger study designed to study psychological aspects of LQTS. Participants were selected from an Internet-based user group that directly serves this specific population. A qualitative research design was utilized within this study to ascertain trends within the user group's message content. This research design was chosen, based on the limited information about this specific population and the ability to understand LQTS from a theoretical framework in order to develop specific themes relevant to this population. The limited psychosocial research on LQTS led to a broad focus on anxiety related themes that can be further investigated in future research. The development of theory from the data will provide the opportunity for future research to analyze specific variables related to these themes.

Design and design justification.

Relatively little research has been conducted on the LQTS population. This syndrome, in comparison with other chronic illnesses, is not conceptually well understood in terms of specific aspects, including medical and psychosocial consequences of the syndrome. In order to become more fully informed about this population, a qualitative design was chosen as a starting point to understand mental health trends (such as anxiety) within this population. This type of analysis provided the opportunity to identify aspects that are specific to this population through gathering descriptive accounts from individuals affected by LQTS. Qualitative analysis examined archival data from a LQTS user group to assess for specific themes related to anxiety.

This user group provided access to a wide range of individuals, including participants of varying cultures, ages, and genders and also allowed access to a group that would be difficult to access in large numbers in other ways (Mann & Stewart, 2000). Limitations of this methodology are discussed in the results section (see Chapter 4).

A qualitative design was chosen for this research to help gain a perspective of individuals' experiences with LQTS and the resulting explanations and meanings that they attribute to this experience. The emergence of specific themes presents essential information to professionals and also serves as an impetus for future research. The development of these themes was based on the concept known as grounded theory.

Grounded theory is the process of building a theory from data obtained from individual perspectives and the themes that emerge within these accounts. Prior to analyzing the data, theories about coping with medical illnesses and anxiety were reviewed to guide the research process. After themes were gleaned from the data, new ideas emerged about a theory that was more directly applicable to the LQTS population (Corbin & Strauss, 2008).

Another aspect of qualitative research that was implemented to ensure a structured process is the use of analytic tools. Analytic tools are strategies that are used during the coding process to ensure an accurate interpretation of the data. The use of these tools ensures that the coding process is an in-depth process that leads to the development of a theory. This in-depth process allows the researcher to understand each piece of data as a single entity and as an integrated piece of data that develops over time. The integration of data in a systematic manner provides the opportunity for the development of a theory that is well formed and is based on significant consideration of all aspects provided by the interpretation of the material (Corbin & Strauss, 2008).

Participants.

Participants were selected from an archived database of individuals on a LQTS user group that consisted of approximately 780 international members. Individuals participating with the user group had been diagnosed with LQTS or had been directly affected by this chronic illness, according to their self-report. This user group allows members to post and correspond through messages related to LQTS, in order to solicit support and information, and social connections.

Inclusion and exclusion justification.

Participants included in the study are those who are directly affected by LQTS whether or not they have the illness or have a close family member with LQTS. Participants are those individuals who belong to the LQTS user group and had posted a message in September 2008 or February 2008. The rationale for selecting these months was to eliminate months that had major holidays or major transitions (e.g. summer vacation) in an effort to avoid the possibility of these factors confounding the data, but also, possibly, to capture seasonal differences. Those individuals who were not included from the user group are individuals who did not respond to messages within the specified time frame or did not recount having anxiety resulting from the illness experience.

Screening procedures determining inclusion and exclusion criteria.

User group messages were distributed to two coding teams. One team consisted of three members and the other team consisted of four members. After messages were coded, the groups met to review their findings. The principal investigator maintained a master copy of all group members' responses to the data that were coded for that week. After reviewing all messages, the researchers reviewed the notes and highlighted the agreement between and among group members. This process is known as triangulation,

and ensures the soundness of results through the agreement of group members. Upon completion of coding of the September and February data, the researchers decided whether or not saturation had been reached at that point. Saturation ensures that no new theoretical constructs could be gleaned from collection of additional data. Additional data were not used because saturation had been established (Marshall & Rossman, 2006). Once saturation had been reached, the process of extrapolating the data began to develop an understanding of anxiety within the LQTS population. This process also took a structured approach to ensure sound theoretical integration (Corbin & Strauss, 2008).

Recruitment.

This study used archival data, thus there was no recruitment of members. Participants responded prior to messages being downloaded for research use. Group moderators gave their permission to the principal investigator of the primary study to use this data after it had been de-identified. The large size of this user group suggested that it was in the public domain and informed consent was not needed from the members (Mann & Stewart, 2000; Eysenback & Wyatt 2002).

Measures.

Based on the qualitative design, there were no measures in the current study.

Procedure.

Initially, user group messages were downloaded into a word document to allow for de-identification of messages and use of search strategies. Messages were analyzed by each of the three or four coders on two separate teams. Collective discussion and analysis took place shortly afterwards to agree on themes. The researchers began conceptualization, based on collective analysis and theoretical groundwork. Those messages coded as anxiety were based on cognitive, affective, physiological, and

behavioral changes; these were self-reported by user group participants and fit the operational definition of anxiety according to the American Psychiatric Association Diagnostic and Statistical Manual of Mental Disorders-Fourth Edition-Text Revision (DSM-IV-TR). Cognitive aspects are related to sensory-perceptual symptoms including: unreality, hypervigilance, thinking problems (such as confusion or poor concentration), fear related beliefs and thoughts (such as fear of losing control or fear of not being able to cope), and depersonalization. Affective changes of anxiety include: intense feelings of guilt, shame, anger, apprehension, worry, being scared, and fear. Physiological changes related to anxiety that results from hyperarousal can include (but are not limited to) sweating, increased heart rate, insomnia, poor appetite, fluctuations in breathing, and dizziness. At the behavioral level, individuals with maladaptive anxiety may exhibit decreased behavioral responses that are related to safety, avoidance, or helplessness (Emilien et al., 2002).

In relation to anxiety, it is important to remember that anxiety can also be adaptive. The adaptive value of anxiety is intended to provoke an intensely unpleasant experience that motivates an individual to take steps to reduce emotions or take preventative measures to reduce the chances of its recurrence. It is suggested that anxiety becomes maladaptive when an individual holds beliefs that lead to the perception of a majority of internal and external events as being threatening (Beck, Emery, & Greenberg, 1985). This broad understanding of anxiety can be further broken down into a categorical approach, based in the DSM-IV-TR. The DSM-IV-TR provides clinicians within the field the opportunity to operate according to a standard criterion for diagnosing mental disorders (American Psychiatric Association, 2000). Based on the scope of this study, it is not possible to diagnose participants based on this categorical approach.

However, it is an important starting point to understand how anxiety may be manifested in this population and also to direct future research. Manuscripts will be given to three moderators of the user group for comment and feedback prior to the submission of the manuscript.

Chapter 4

Results

Data for this study were collected from a LQTS user group consisting of approximately 780 international members in the months of September 2008 and February 2008. From the month of February, data analysis identified 298 posts within 36 threads. Within these threads, 58 different individuals participated by posting to the LQTS user group. In addition, 26 of the participants had a child with LQTS; 4 had a sibling(s) with the diagnosis, and 2 had a spouse with LQTS. From the month of September, data analysis identified 413 posts within 44 threads. Within these threads, 90 different individuals participated by posting to the LQTS user group. Of these individuals, it was indicated that 38 had LQTS. In addition, 41 of them had a child with LQTS; 8 of the participants had a sibling(s) with the diagnosis, and 4 participants had a spouse with LQTS. Participants were not asked directly to disclose information; rather, information was gathered through the unsolicited postings by participants. Based on this method of information gathering, more individuals could have been or are affected by LQTS; however, this gives a general representation of individuals within the user group.

Further analysis of the data in February indicated 53 of the 298 posts (18%) were coded as representing anxiety by a coding team of four members. Within these 53 posts from February, 33 different individuals responded in the message threads. Of these 33 individuals, 10 identified themselves as having LQTS; 13 identified their child(ren) as having LQTS; in addition, 1 parent identified a sibling; 4 identified both themselves and child(ren) as having LQTS; 3 identified themselves and their siblings with the diagnosis; 2 members identified an extended family member; 2 reported their husbands and child(ren) had LQTS as well as one identifying extended family members; 1 member did

not provide any information about specific individual with LQTS. Individuals included in the month of February self-reported their locations in but not limited to South Africa, New Zealand, Scotland, and United States.

Further analysis of September data indicated 69 of the 413 posts (17%) were marked as presenting anxiety by a separate coding team of three members. Within these 69 posts, 27 individuals responded and 7 of these individuals also responded to posts in February. Of these 27 individuals 5 described themselves as having LQTS; 7 identified their child(ren) as having LQTS; 4 indicated that both they and their child(ren) have LQTS; 2 identified themselves and their siblings as having LQTS, and 2 identified their child(ren) and siblings as having LQTS. Additional analysis indicated that 1 individual identified herself, her sibling, and an extended family member as having LQTS; 1 individual reported her husband and children as having LQTS; 1 individual indicated herself and her grandchild as having LQTS, and 4 individuals provided no identifying information. Within the months of February and September, fourteen messages were excluded because the contents of their messages were outside the scope of the study

Based on the analysis of the data, information about two significant time periods were gleaned from discussions among participants in the user group as having rendered different experiences. These time periods suggested within the message contents are based on length of diagnosis, which include the initial diagnostic phase and continued treatment experiences related to LQTS. Within these two time periods, there were three identified themes: “Uncertainty”, “Unpredictability”, and “Uncontrollability”.

Initial treatment of LQTS.

The first time period reflects information about those individuals who recently have been or have had a family member diagnosed with LQTS. Throughout these

messages there appear to be three separate themes: uncertainty, unpredictability, and uncontrollability.

Uncertainty.

The first theme that developed within the data was uncertainty, and this is suggested to contribute to anxiety within participants. Based on the data, this uncertainty appears to be related to two different experiences discussed by participants.

What syndrome.

When individuals were first diagnosed, they expressed a lack of certainty due to an unclear diagnosis and to direction regarding medical recommendations. Numerous participants discussed situations directly related to the diagnosis of LQTS that create a sense of uncertainty, leading to feelings of anxiety. Several participants expressed the ambiguity that occurred during initial stages regarding their diagnosis. A participant explained that her daughter was referred to a specialist for a borderline diagnosis, but was medically cleared. This participant stated “I think that I will always wonder and even worry since she has only had two EKG(s) and no other test if I don’t get another opinion.” The participant received two different results from physicians that led to feelings of uncertainty and anxiety about not treating her daughter. Another participant explained her experiences leading up to a more certain diagnosis. She stated that in the past medical professionals had medically cleared her and told her not to worry about her symptoms. She explained how she continued to endure these symptoms until a particularly significant cardiac event. She explained, “I know this is a stupid question, but should I be concerned? I really don’t know what to do next. The doctors gave me no resources or information...nothing that I could count on for the future, no further tests or medications (one of the reasons I don’t have much trust in my doctors).” Another mother

confirmed these uncertainties by stating, “Thirdly, without being able to pinpoint the exact type of LQT that Mark* (name changed for reasons of confidentiality) has, we are left treating all of the known triggers for episodes. Should we take alarm clocks and phones out of our house? Should he have a sleep monitor? Should we let him swim? Let him play sports? Do you limit everything or nothing?”

Throughout the data, participants describe attempts to cope with anxiety through different means. One mother stated, “She said I have LQTS with a mutation on the KCNE1 gene. Then told me I need to get my daughters tested and probably my brother and that she was sending a letter explaining this. End of conversation...My EP seems to think this is all I need to know unless she has written more in her letter.” Similar to other participants, this mother sought out support and information from the user group to help cope with this anxiety. It has been observed many times in this community that participants are able to offer support to a certain extent; however, there is still a pervasive lack of knowledge related to this syndrome resulting in continued anxiety.

Waiting game.

Participants also discussed a lack of certainty about the course of treatment within the LQTS community. One member stated, “At the moment I am pretty much confused. The doc only said that there is no change in meds-we are waiting for more explanations and direction in the coming weeks”. Another individual stated, “Can you give us advice on what to expect now? Are these results ok or do you think we should be worried[?] We have also been advised to stop him from doing any sports until we have been seen next week[.] Is this normal as well; we would like any feedback as we are unsure of what we are dealing with, thanks.” The uncertainty that many participants expressed result from the patients’ perception that the information received from their doctors was vague

or unclear. One participant expressed this experience of uncertainty after initially being diagnosed, including her difficulty in determining a definite course of treatment because of the perceived vagueness regarding medical recommendations. This specific participant explained that her doctor permitted her to continue with her current, physically active lifestyle even though she is suspected to have LQTS. She asked the group for advice because of the vagueness of the diagnosis and stated, “My doctor told me that for this month I could keep playing rec(reation) sports and running to ‘keep up my fitness’ (I run half marathons) as long as I don’t push myself, but now I am a little worried that something bad might happen.” It is suggested that many times individuals in these initial stages are looking for clear information about how they should approach treatment. Many times it appears to participants that medical professionals present vague information, such as the situation previously mentioned. This specific participant was told, “Not to push herself”, yet her definition of exerting herself physically may be completely different from that of an individual who does not run marathons.

Within this subtheme, participants are again seen making an effort to cope with this syndrome through seeking out support. An individual explained her initial diagnosis experience, and now is looking for more answers related to her treatment. In addition, she provided a summary statement about uncertainty, which seems to trigger anxiety for many people on the user group with LQTS. She stated: “I feel I’m getting mixed signals on the seriousness of this condition. My confusion. My cardiologist would have done nothing except monitor and I feel it needs more attention now.” Another participant spoke of active coping efforts and attempting to alleviate anxiety through seeking out medical journals. She stated, “I am truly just looking for answers as I am barely 1 year into my diagnosis. As I am an engineer I would like to think I am fairly intelligent, but I

am getting more and more confused about LQTS the more I read. Perhaps I should stay away from reading all of the studies since I have zero medical knowledge!”

Interestingly, this person’s attempts to problem solve and cope with anxiety was further exacerbated by the information she sought out about the syndrome.

A developing trend appears to be that individuals cope actively through similar methods such as seeking out support or reading literature. These coping efforts may stem from the perception that medical professionals do present information about how to manage LQTS effectively in a vague manner. Thus this vagueness leads to active coping efforts that further exacerbate anxiety for two identified reasons. First, other individuals within the LQTS community experience similar interactions with their doctors and have similar concerns and questions. Second, information gathered by participants in an effort to cope effectively is difficult to understand.

Unpredictability.

The second theme is related to the unpredictability of the syndrome and is suggested to contribute to anxiety within participants. Participants discussed two common experiences related to unpredictability in the initial diagnostic phases.

Big changes, really quickly.

Many participants express hesitation about major lifestyle changes because of what seemed to be lack of awareness about the serious nature of the syndrome. The unpredictability of LQTS is unknown to those recently diagnosed with LQTS. For instance, one participant asked, “Are these results ok or do you think we should be worried[?] [W]e have also been advised to stop him from doing any sports until we have been seen next week[.] Is this normal[?] [W]e would like any feedback as we are unsure what we are dealing with [currently].” Family members often view their child or relative

as being healthy, and do not recognize the need to make lifestyle modifications at that current time.

A common trend is for participants of the user group to emphasize the unpredictability and serious nature of the syndrome in order to evoke an anxiety response in others; this is done in an effort to get them to act in a way that is more conscientious and responsive to the diagnosis of LQTS. For instance, one participant responded to another's concerns about modification by stating, "Not to scare you but to encourage you to use caution I will tell you about my uncle who ran 10 miles every day. He was in his 50s and just dropped due to cardiac arrest." In addition, this participant explains, "It can be scary. I have lost others in my family who were healthy due to this."

It's serious, so what do i do now.

Once the unpredictability of the syndrome is understood, people seem to have difficulty negotiating the best course of action. One individual explained, "This has been a real roller coaster ride for me and my family. I feel like I'm living my life on eggshells right now and if one breaks I will die. I am being monitored closely, but unless I have a "life threatening" episode, I have to wait for the scheduled dates." Another participant spoke of her anxiety because she understood the serious nature of the syndrome, but has limited options about how to cope effectively with this anxiety. She stated, "The frustrating thing is that I've been told I'm fine and believed it through the pain for years. Now, I don't know what to think, because all I can remember is that diagnosis and then being told it wasn't something to be concerned about, when there are so many young people (and older) who have died suddenly from a cardiac arrest." Another participant shared similar anxiety about the unpredictability of the syndrome. She explained her family history to add context to her concern and stated, "My mother and brother both

passed away in their sleep and my sister passed away after running to get a ball in the driveway. I am getting an ICD next week and have appointments set up for both of my children. They are 15 and 11 and I am terrified...I am trying to learn more about the best way to protect them without prohibiting their life.”

Uncontrollability.

The third theme that developed within the data is uncontrollability and this is suggested to contribute to anxiety within individuals. Based on the data, participants discussed two experiences that contributed to feeling a lack of control.

Learning to walk again.

People with LQTS feel a lack of control in relation to this syndrome. Lack of control may relate to their experiences of anxiety. LQTS influences individuals' lives after they have led typical lifestyles, involving little expectation of having a chronic illness and the associated life altering consequences. One family described their active lifestyles with no suspicions of LQTS until the sudden death of their younger child. In a response to another parent, the mother stated, “Emotionally you were hit by a Mack truck when he was born and now you are still learning to walk.” This mother clearly identified the loss of control in many aspects of life related to this syndrome, including how one has to try to make modifications and cope in order to gain some semblance of control over one's life. Other participants express the loss of control and the feelings they experience as they are waiting for answers and direction. One individual stated “I know it will seem like a long time till Monday, but hopefully you will have some answers by then.” Similar to this individual, another individual expressed her thoughts, “I find right now that the worst part does just not know what is going on.”

Participants also speak about their coping efforts related to regaining some sense of control over the situation. One new mother explained, “But it’s my baby I’m so worried (a) bout; she’s 9 months now and is doing so well but I’m so worried that something will happen to her; she hasn’t had any episodes and I’m hoping she won’t but I’m constant(ly) watching her when she’s asleep and awake and if she even takes a deep breath I go weak at the knees...” Similar to this mother, others express the lack of control they feel related to LQTS and seek out information from others to try to cope with their anxieties. One mother asked, “Now you need to let us know, how you watch her every day? How do you travel or go out? Do you take your AED every time with you? Did she have any attacks/fainting again? How safe is to keep her on Beta Blockers for another 3-4 years before ICD?” The questions from participants are numerous, in an effort to gain some semblance of control over their lives and over LQTS. Many times, these questions are not answered and it is suggested that they exacerbate anxiety responses in participants.

On top of everything else.

Participants expressed feeling a lack of control related to extraneous aspects of LQTS. One extended family member explained how she had been diagnosed with LQTS and had anxiety, which was related to other family members not being tested. She stated, “I just hope they don’t wait too long. This really adds to my stress but I keep telling myself that it is not my fault and I can’t really control what she does with her family...right.” Another participant spoke about other factors that led to feeling a lack of control related to LQTS. She discussed aspects of insurance and medical coverage and the lack of control that is felt by many participants related to paying for LQTS medical procedures. At one point she stated, “Many people have found that persistence is

required to get the insurance company to cover genetic testing”. Another participant spoke about insurance coverage and explained her current situation, “Yesterday afternoon, I received a phone call from the EP office saying the insurance company was refusing to give pre-authorization for it (ICD placement) as they were determining it was not medically necessary.”

Continued treatment of LQTS.

The second time period reflects the experiences of those individuals who are past the initial diagnosis of LQTS and have had several specific experiences related directly to LQTS. It is suggested that these themes fall into the same three separate categories mentioned beforehand, and are further differentiated into experiences that are suggested to contribute to anxiety in participants.

Uncertainty.

Within this second time period, the first theme that developed was an uncertainty, suggested to contribute to anxiety within individuals. Based on the data, participants discussed two experiences that contribute to feelings of uncertainty.

What if's of medical procedures.

Participants expressed concerns about medical procedures related to LQTS and the lack of understanding regarding these procedures. Several participants relayed the uncertainty they experience related to medical procedures. One mother expressed her concern about her young daughter receiving an ICD and stated, “Her doctor is starting to talk ICD...☹...much as I hate the idea. Surgery is surgery, and an ICD is not something you dispose with one day.” It appears that often with these medical procedures, the uncertainty and resultant anxiety is produced not only by the procedure itself, but also the doctor's apparent lack of certainty in the diagnosis and course of treatment. One

participant explained her feelings of anxiety by stating, “But, I don’t have a lot of confidence with a civilian cardiologist either seeing as how the first cardiologist that I saw was a civilian and he could ‘neither confirm nor deny’ that I had LQT.” Another mother stated, “Well guys Sally’s* surgery is in the morning. I’m scared. Really really scared. Dr. Jones* says that they’ll have everything set up “just in case”. I don’t know if I mentioned it before but, Dr. Jones* also said that Sally’s* original diagnosis of LQT3 is wrong. He is not 100% sure what it is[,] he does know its LQT oh and I found out her interval runs around 515 and higher....sooo anyways, he said that she most likely has a mutation that hasn’t been found yet. Not a real big relief for me I suppose” (names changed for confidentiality purposes). A third participant expressed further anxiety felt by many participants, asking several questions about her surgery and stating: “Sorry for drilling you [,] but I am waiting to have an ICD implanted and I am getting nervous about the whole thing.”

Often, participants express the idea that failure to receive solid feedback about the correct course of action exacerbates this uncertainty. One mother encapsulated this theme by stating, “We can also drive ourselves crazy with worry and the What if’s.” She further emphasized, “It is difficult to find that balance between reacting and over reacting.” One father stated, “I really never know what right thing to do is and my wife and I fall out over it. Now the next big thing is are we doing the right thing by getting him [an] ICD when he has been fine.” The father added further evidence by stating, “The other thing [is] we only see [a] consultant like every four months unless something happens; our local doctor cannot really advise [us] as [they] know nothing about [the] condition so [deleted] it’s like we wait for something to happen next before we move on to the next stage...that’s why [it] feels like we [are] waiting for something to happen..?!”

Another participant stated, “Doctors look at all kinds of factors-family history, qt interval, history of symptoms, etc-and try to assess your risks and how well the medicines are working for you, but it’s not an exact science. Sometimes different doctors have different opinions for the same case.” These experiences are pervasive within the online community, although they occur in many different medical settings. One mother explained a recent trip to an emergency room and stated, “I was grateful for this because at the time of it all happening didn’t think the emergency doctors took us seriously. They knew about Jordan* having LQT, but put the seizures down to her hitting her face on the deck” (name changed for confidentiality purposes).

Future concerns.

Individuals expressed future concerns that exacerbate anxiety. Based on participants’ responses, it is suggested that the lack of clarity concerning this syndrome appears to produce uncertainty regarding the future. Participants expressed concern about the continued uncertainty related to the syndrome. One participant stated, “One of my fears used to be, “what if I pass out at home.”

Other individuals indicate similar experiences of uncertainty that lead to feelings of anxiety because of not knowing the answers to experiences directly related to LQTS. One participant stated, “...actually not sure what it was, but I suspect it might have been my ICD.” In addition, the uncertainty often continues because of a lack of support from the medical community; he stated, “And then I didn’t want to call my doc about this since she’s always so very busy and hasn’t even time to write the usual reports and hasn’t written one since over a year.” Another participant adds evidence to the continued uncertainty experienced by individuals affected by LQTS and stated, “If you are really nervous, be sure to indicate that to your EP. I know my EP knows I am a fruitcake when

it comes to surgery. Although I've "been there, done that" several times, it doesn't make the idea that much easier." Interestingly, one individual provided further evidence about the general lack of support in the LQTS professional community that creates anxiety about the future by comparing their own experience with that of the cancer community. She expressed this by stating, "The wonderful thing is that, unlike LQTS, there is SOOO much support and research out there for breast cancer".

This subtheme continues to provide evidence of cognitive aspects of anxiety. These participants express the nervousness and worry they feel towards these situations. One participant explained her emotional reactions as "being a fruitcake." Similar to other subthemes, participants are seen trying to cope adaptively with their anxiety; however, based on prior experiences, they are hesitant to engage in adaptive behavioral responses because of these prior experiences. Thus they continue to experience anxiety and do not cope adaptively or resolve their anxiety because of perceived barriers.

Unpredictability.

The second theme that developed within the data is unpredictability, which is suggested to contribute to anxiety within individuals. Based on the data, participants discussed three experiences related to unpredictability in the second time period.

Silent striker.

Participants often feel and appear healthy to others, but always have a lingering anxiety because of previous experiences with this syndrome and its unpredictability. Participants describe a sense of not knowing what to expect from one day to the next. One participant encapsulates this idea when expressing the urgency needed in regard to treating this life-threatening syndrome. She stated, "It always bothers me when I hear the "you've lived this long with it..." line. Yes, my mother passed out many times over her

lifetime. She was never diagnosed or treated for long qt. She died from it at age 57. Her retrieved EKG's from 14 years earlier showed a long qt interval. Too darn bad this got buried in her medical records and the right person never took it further. Please, make sure you get treated for long qt, even if you've never had symptoms."

It is suggested that part of the difficulties experienced by these participants is how "normal" they feel and appear to others. One participant sums this up by stating, "I think it is the unpredictability that makes LQT so hard to adjust to. We usually look perfectly healthy, we might have no symptoms at all, but there's no way of knowing if or when we will (or how severe)". One participant explained the unpredictability of the syndrome by stating, "Just when we were getting used to our diagnoses and life was starting to get back to normal WHAM!" Another participant added further evidence of this unpredictability in response to this participant by stating, "We get used to "normal" and then all of a sudden the "silent striker" gets us" and "Long QT is strange in that it can give us one really bad day in the middle of so many great ones. But the fear does linger around for quite some time."

Parental concerns.

Several parents of children with LQTS reported anxiety directly related to concerns about their children. One participant explained that after a seemingly normal day, she got a phone call that her son was taken to the hospital. She stated, "My head is just spinning and I feel totally out of it." Another participant added additional information about the unpredictability experienced by families with LQTS and stated, "Anyway, the phone call you received from George's* school is our worst nightmare, a call that we all know CAN happen but one we certainly wish never happens to any of us."

and “We try so hard to protect them, knowing all the while we cannot protect them from everything!”

Other parents speak of how they handle the unpredictability. Many participants expressed perceived safety in the ICD. One participant stated “The best thing about an ICD is for the confidence in the “protection” when we can’t be there. Fred* had only one shock and that was at a time when he was alone. I don’t even want to think what could have happened if it wasn’t for his ICD. Although, the ICD is not without problems, it gives protection we otherwise wouldn’t have.” Another mother discussed her coping efforts to handle the lack of predictability related to LQTS. She explained, “Since she is so young, she is never out of our sight. So we can, for now, provide protection for her with the AED since she doesn’t spend any time alone.” This coping mechanism emphasized strategies that the family used to handle the unpredictability of this syndrome. It is suggested that these coping efforts only decrease anxiety to a certain extent, based on participant’s statements regarding LQTS being a constant threat, no matter what course of action is taken to cope with this syndrome.

How many.

Participants’ perceptions of the number of triggers that can contribute to a cardiac event further increased perceptions about the unpredictability of the syndrome. One participant presented the idea of unpredictability of LQTS by talking about the numerous triggers to cardiac events and stated, “This just goes to show that LQTS is so unpredictable.” Another person stated, “The odd thing is that since she went onto betas and I’ve managed her life (hydration, proper eating, potassium, etc.); she hasn’t even had so much as one dizzy spell.” One mother stated, “We are grateful we understand her trigger. However, we are not naive enough to think that she could not have a cardiac

arrest due to another trigger. All of her episodes have been at rest or while she is sleeping. That is why we sleep with her between my husband and me.”

The unpredictability corresponding with the diagnosis of LQTS may also lead participants to engage in several different types of coping responses; this speaks to individuals’ perceptions of the number of LQTS triggers, and how it is unpredictable it is, and whether or not an event will occur because of these triggers. One participant explained her perception of things by stating, “I’m glad to hear you are going to get back into things and haven’t given up due to the fear of the shock. I hear a lot of people after a shocking storm like yours withdraw from activities and become scared to sleep, etc., always fearing the shock.”

Similar to other participants included within this subtheme, others provided additional evidence of the unpredictability by stating how certain things help them cope with these feelings of anxiety. In addition, another participant stated, “It is such a relief to know that my ICD will give me the protection when I need it and to know that our EP is able to do the procedure”. Another individual indicated a cognitive technique that she has used to cope with unpredictability of the syndrome that helps ease her anxiety. She stated, “It seems like the most treatment complications can be dealt with. I usually cope by thinking of the worst that can happen. Then I think of how I would deal with that. Usually I can figure out how I would and it isn’t as bad as the anxiety about the unknown.”

Uncontrollability.

The third theme that developed within the data is uncontrollability and is suggested to contribute to anxiety within individuals. Participants discussed two experiences related to uncontrollability and anxiety.

No voice.

A participants' feeling of a loss of control in making medical decisions leads to feelings of anxiety. One participant stated, "They monitored him at the hospital for a couple of days and things started to normalize (as normal as it gets)." This participant alludes to the fact that often there is a loss of normalcy and control over one's life when dealing with this syndrome. Another participant wrote in response to this situation with a participant's child, "I think the thing that makes it even worse is not having control over what's going on," and "I know these are only a drop in the bucket of all the worries you're feeling, but maybe even a little thing will help him feel better." This participant alluded to the idea of a "bucket of worries" similar to the way another participant explained her experience with LQTS. She stated, "OK, I'm fine with having long qt. No problem with the icd...Can handle the fatigue, medications, etc. BUT, what I really cannot understand or really ever grasp is the stress caused by the doctor/insurance political absurdity."

Some participants allude to the fact that their lack of control has led to problematic anxiety responses. One participant indicated that she has been diagnosed with anxiety disorders and stated, "I have also been diagnosed with PTSD and also have extreme anxiety. I am much better now but my biggest problem is going out alone. It scares me in a situation to not know someone." These anxiety responses occurred after several shocks that continued to occur because the hospital staff inappropriately handled the situation. Another participant shared a similar story about traumatic experience within a hospital that led to multiple shocks, loss of blood, and emergency surgery. This individual stated, "Since then I've been dealing with anxiety issues and seeing a psychiatrist for post-traumatic stress disorder."

Support.

A poor sense of control is related to the perceived low support within participants' actual communities; this is in addition to feeling as though there are limited resources that could otherwise help participants gain a sense of mastery over this syndrome. One mother spoke about a birthday party and how her plan to keep her daughter safe met with an unfavorable response from the mother hosting the party. Although this mother is attempting to alleviate her anxieties about the situation, others not viewing it in the same light make it difficult. In addition, participants also suggest a lack of control related to information. One mother stated, "My daughter (15) was diagnosed with LQT5 almost 2 years ago now and ever since then I have been trying to gather as much info as I could on it-I'm beginning to think it doesn't exist". In a sharp comparison, another participant speaks of her experience with LQTS and cancer. She explained that unlike LQTS, "I found all the "options" and decisions extremely overwhelming, but it did help me to feel in control of my own care to be involved." Many of the participants speak of wanting to be in greater control of this syndrome, but lack the options that this person expresses that create a feeling of comfort in relation to her care and overall well-being.

Conclusion.

Based on the results, there are several suggested experiences that lead to anxiety within this population. The inherent nature of LQTS leads to a certain level of uncertainty, unpredictability, and uncontrollability. It is suggested that these individuals have difficulty coping with LQTS because of the lack of support and information within the community. This lack of support and information within the community may contribute identified obstacles and barriers to cope effectively at times with this syndrome, and this is suggested to lead to anxiety. Based on these initial findings, it

becomes evident that experiences unique to this syndrome could lead to maladaptive anxiety responses. Based on the data and overarching conceptualization of anxiety, the discussion will address the specific themes describing the experiences of participants that may have led to anxiety responses. In addition, a proposed trajectory could be hypothesized about how maladaptive anxiety could develop within this population; this will be further expanded upon in the discussion.

Chapter 5

Discussion

Analysis of data revealed several themes in relation to anxiety and LQTS. There appear to be two time periods, initial diagnosis and later treatment experiences, in which anxiety is experienced in participants. During these discrete time periods, participants expressed anxiety related to experiences that created uncertainty, unpredictability, and uncontrollability. Themes were developed out of these three areas, based on shared experiences by participants. To further elaborate upon the findings, the application of the findings into a cognitive behavioral context will help elucidate how anxiety could become maladaptive within the LQTS population.

Development of anxiety and implications to LQTS.

Anxiety is an emotional state that is stimulated when an individual experiences fear. It is an adaptive response that directs attention to a stimulus within the environment. In an effort to decrease anxiety, an individual will engage in strategies or coping responses. Anxiety becomes maladaptive when the person continues to experience anxiety that impacts their overall functioning due to a misperception or exaggeration of danger (Beck, Emery, & Greenberg, 1985). Maladaptive anxiety responses impact one's function in social, occupational, and other areas (DiTomasso & Gosch, 2002).

The data clearly identified triggering factors that contributed to anxiety within this population. Based on the nature of the study, it was not possible to differentiate whether anxiety responses in participants were adaptive or maladaptive. When considering whether the anxiety an individual is experiencing is adaptive or maladaptive, it is helpful to look at the concept of maintenance of anxiety. Individuals who experience heightened

anxiety and have poor coping skills often engage in behaviors that do not lead to resolution of the anxiety. These behaviors are escape and avoidance behaviors that lead to the removal of the anxiety-provoking stimulus. However, the individual never learns there is nothing to be feared regarding that particular stimulus (DiTomasso & Gosch, 2002). The maintenance of anxiety and how it could become maladaptive within this population will be discussed within the cognitive behavioral framework of anxiety.

Cognitive behavioral model of anxiety.

Chronic illness affects every aspect of an individual's life. The extensive nature of chronic illness pervades one's social, academic, medical, and personal life realms. A thorough understanding of how chronic illnesses affect these aspects is necessary, as also are the resulting psychosocial consequences of required lifestyle modifications. Multiple aspects of chronic illness are not controlled by an individual and increases susceptibility to anxiety and other psychosocial effects. The data showed that participants were experiencing anxiety reactions to triggers within the environment.

It is helpful to look at some of the assumptions of the cognitive behavioral model to start to discern the anxiety experienced within this population and whether it is adaptive or maladaptive. The main premise of all anxiety responses is that fear is an adaptive response that is essential to survival. Anxiety becomes maladaptive when there is misperception that leads to an overestimation or exaggeration of fear. Within a cognitive behavioral context, cognitive, physiological, motivational, and behavioral systems play a vital role in anxiety. However, the cognitive system is viewed as primary to elicit anxiety reactions because the cognitive system appraises the danger and the perceived resources that an individual has in order to handle the stressor (DiTomasso, Freeman, Carvajal, & Zahn, 2009).

These beginning elements of the cognitive behavioral model display how maladaptive anxiety develops. In the current research data, it is evident that there are specific stimuli/stressors that elicit a fear response in participants. Based on the data, it is suggested that without further research one cannot make the assumption that these perceptions of fear are maladaptive. Although there are several triggers identified within the data, a common theme that appears throughout the data is that individuals perceive that they lack the ability to cope effectively because of scarcity of resources and information. Without further data, it cannot be determined whether or not these are cognitive misappraisals that lead to maladaptive anxiety responses.

The cognitive behavioral model further breaks down the development of anxiety. Over time, it is suggested that these misperceptions lead to errors in thinking (known as cognitive distortions) that are fueled by beliefs systems within an individual. These thoughts and beliefs influence those stimuli that an individual is attuned to within his or her environment, leading an individual to misinterpret ambiguous stimuli as threatening, thus creating anxiety (DiTomasso, Freeman, Carvajal, & Zahn, 2009). These beliefs tend to be rigid, over generalized, and global, which can present problems for an individual if they become skewed in some way to distort an accurate interpretation of events (Beck, 1995).

Another way to expand the idea of belief systems specific to chronic illness is illness representations. Personal models of illness or illness representations are specific to illness related stressors and consequences. This idea is based on the concept of schema but directly relates to illness. Personal models influence emotional and behavioral responses to illness stressors. These personal models incorporate past beliefs with recent information. In addition, personal models incorporate emotional aspects of illness that

have been neglected in previous health belief models. These models examine the variables that influence the overall experience of illness, management, and emotions related to the illness. It has been suggested that personal models have five components: identity, cause, timeline, consequences, and control (Skinner & Hampson, 2001).

The first factor, identity, defines specific labels and factors associated with the illness, such as shortness of breath, fainting, and lightheadedness (Edgar & Skinner, 2003). The development of a belief system related to LQTS starts with this specific component. Throughout the data, participants express anxiety related to the number of triggers that could cause a life threatening cardiac event. Unlike other chronic illnesses, participants affected by this syndrome have another conflicting factor when trying to define the identity of this syndrome accurately. Participants consistently express a desire to know what specifically will predispose himself (or herself or a family member) to a cardiac event and what types of symptoms they will experience during an event. It has been suggested that participants develop anxiety over the inability to define, with accuracy, this syndrome that has altered their lives.

The second factor, causes, are beliefs about what specifically has caused the disease and the person's role in the onset (Edgar & Skinner, 2003). This component of personal illness models was not further delineated within the data. It could be speculated, however, that further research regarding parental perspectives revolving around the cause of this syndrome will provide interesting research outcomes.

The next factor, timeline, indicates the estimated length of time and the symptomology that will be recurring (Edgar & Skinner, 2003). This component is another dynamic factor related to LQTS. First, many participants unexpectedly are introduced to LQTS by being diagnosed themselves or by having a family member

diagnosed. These individuals are asked to make significant lifestyles changes in a short time, although they have felt completely healthy for most of their lives. It is suggested that the lifestyle changes create anxiety when a medical professional is unable to give definite answers about whether or not they will ever have symptoms, or what type of symptoms they will experience, related to LQTS. A second area of LQTS related to the timeline component is the time at which participants recognize the severity of the syndrome. Medical treatment or recommendations are often delayed due to the need for further testing, referrals and so forth. The lack of certainty while waiting for a diagnosis and for medical recommendations will probably continue to evoke anxiety responses. In addition, during this period of waiting, participants are unsure of how to keep themselves or a family member safe from having a cardiac event. The third area related to timeline is the unpredictability of the syndrome, which includes not knowing when to expect symptoms. In addition, participants express anxiety because they have no ability to control these symptoms, thus dismantling the ability to manage and predict future events related to LQTS.

The fourth factor, consequences, are beliefs about psychological, social, physical, and other aspects expected to occur from the illness (Edgar & Skinner, 2003). Perceived consequences of the illness are similar to perceived susceptibility and severity of the health belief model. It has been suggested that these perceptions include the beliefs that are most influential in relation to emotional consequences (Skinner & Hampson, 2001). This specific component of a personal model of illness has several implications for the LQTS population. First, participants express anxiety over specific medical procedures that occur, related to this syndrome. Second, the unpredictability inherent in this syndrome leads to a poor understanding of the possible consequences of this syndrome.

This unpredictability is suggested to create anxiety because of the inability to clearly define consequences related to this syndrome. The third area related to LQTS and its consequences regard the uncertainty that participants perceive regarding the overall implications of the syndrome and course of treatment. Participants clearly expressed continued uncertainty regarding consequences and ways to manage LQTS effectively. Participants describe desiring clarity from medical professionals about how to manage LQTS; instead, they feel as though there is continuous uncertainty regarding LQTS and resultant anxiety.

The last factor, control, relates to treatment effectiveness and the overall implications of the illness in the short and long term (Edgar & Skinner, 2003). Through the data, it became evident that participants feel a loss of control related to this syndrome. Participants consistently disclose the fact that they feel a lack of control about effectively handling this syndrome. Many participants express the idea that this lack of control is related to their perceptions related to interactions within the medical setting. In addition, participants express a lack of control because of their perceived low support within other social communities; others view them as healthy. Beyond that, there is a lack of awareness within the general public about LQTS.

The concept of schemas or personal models of illness is an interesting concept related to this population. From a cognitive behavioral perspective, the premise of schemas includes danger and threat beliefs that lead to anxiety reactions. The problematic response arises when an individual starts to have limited objectivity that leads to selective attention to stimuli in his or her environment rather than to a realistic, rational appraisal of the situation (DiTomasso, Freeman, Carvajal, & Zahn, 2009). Based on data realized from this research, it is suggested that the continual themes involving

lack of information and lack of clarity are adaptive, realistic triggers for anxiety. Many individuals affected by LQTS cope effectively with their anxieties. However, it is suggested that continued experiences related to LQTS lead to the possibility of individuals developing belief systems related to harm and vulnerability, ultimately leading to the susceptibility of developing maladaptive anxiety in the future. With this suggestion, it becomes imperative to continue to develop further research with specific data identified within this study that could lead to a comprehensive understanding of maladaptive anxiety within this population. In addition, taking active steps to help participants cope with the inherent unpredictability, uncertainty, and uncontrollability of LQTS would decrease their suggested adaptive levels of anxiety.

Biopsychosocial model.

Aspects of LQTS can be anxiety provoking, which suggests the need to take active steps to help individuals affected by this syndrome cope. Many participants in the user group expressed a desire to cope adaptively with this syndrome, yet they lacked the support and direction to do so. It has been suggested that the major aspects of LQTS that are highly influential in contributing to participants' anxiety are the lack of support and information from the medical community; these aspects are influenced by the model that has been adopted by the medical community.

The biomedical model has been the primary model utilized within the medical community. This model focuses primarily on disease and disability. It examines the causes, prevention, and cure of disease; however, it has been suggested to have several limitations. The most highly recognized limitation is that it does not incorporate psychological, social, and environmental factors that influence the disease process. This model continues to appear to be dominant within the medical community. It has been

suggested this dominance is a result of medical training and experiences that lead the medical community to recognize physical signs and symptoms, and not recognize psychosocial influences on disease. With increased knowledge of psychosocial influences on disease, there has been an increased recognition in the medical field of the need to incorporate other factors into their understanding of the disease process (Sperry, 2006).

The psychosocial model examines how an individual copes with stressors directly linked to the chronic illness or disease. This model is heavily influenced by Lazarus and Folkman's theory of stress and coping. As discussed previously, cognitive appraisals of harm and threat influence how an individual copes with a stressor. This model adds the effect of coping to disease related behaviors. However, it continues to be ineffective in ways in which it can affect one's overall health functioning (Sperry, 2006).

The biopsychosocial model is the most broad and integrative of all the models. This model encompasses biological and psychosocial factors of chronic illness. This model recognizes how the biological function of illness, stressors, and coping interact in order to lead to disease progression and associated illness behaviors. This model further expands upon the concept of stress through integrating psychological, physiological, and environmental levels that affect an individual's overall functioning (Sperry, 2006).

Overall, it is suggested that the anxiety experienced by this population is a result of difficulty in coping with this syndrome; this is a result of lack of clarity and lack of information about how to handle LQTS effectively. It is suggested that creating a biopsychosocial approach to care will lead to a significant decrease in anxiety experienced by those affected by LQTS. The recognition of psychosocial factors specifically related to this syndrome is essential in order to incorporate this model into

the medical setting. It is also suggested that these individuals will appraise situations as more of a challenge because they will have the needed information and support to overcome or effectively handle their anxiety.

Role of clinical health psychologist.

Through this research, there comes recognition of the need for the incorporation of the biopsychosocial model into the medical setting. The integration of this model would lead to a greater understanding of the psychosocial aspects that account for quality of life in this specific population. The role of a clinical health psychologist as a behavioral health consultant would greatly aid this process. A clinical health psychologist has the ability to provide specific information and recommendations related to individuals' behavioral, emotional, and overall psychosocial functioning (Gatchel & Oordt, 2004).

There are many needs within the LQTS population that could be addressed by a clinical health psychologist. On a more general level, a clinical health psychologist could provide general psychoeducation related to typical emotional responses to a diagnosis of a chronic illness and could address ways in which to address these emotional responses effectively. In addition, education focused on symptomology (i.e. hopelessness, loss of interest) that could be evidence of a patient experiencing psychological distress may be helpful. A clinical health psychologist could also provide education surrounding typical psychiatric diagnoses that are present in chronic illness populations. Last, the clinical health psychologist could help medical facilities implement specific protocols to address psychosocial aspects. For instance, a clinical health psychologist could suggest a measure that specifically targets anxiety and/or depressive symptomology and educate professionals about how to utilize this instrument. In addition, it may be useful for a

psychologist to discuss the referral process and the times when referrals should be made for follow up services. Ideally, the clinical health psychologist could directly provide screening and triage services.

More specifically, within the LQTS population, a clinical health psychologist has the potential to make a significant impact on LQTS individuals' psychosocial functioning. Clinical health psychologists should be aware of the significant psychosocial implications of this syndrome due to the inherent uncertainty, unpredictability, and uncontrollability of this syndrome. Based on individual evaluation, recommendations can be made accordingly; these might minimize the impact of the changes required or might help patients and their families cope more effectively.

Future research.

Psychosocial research on LQTS is a relatively understudied field, yet it affects the lives of many individuals. This research sought initially to define, broadly, the anxiety experienced by individuals affected by LQTS. Although this research was able to begin narrowing the specific concerns related to this study, there is still much to be understood, relative to anxiety within this population. The inherent nature of this syndrome leads to vulnerability. In future research, it would be important to further delineate adaptive and maladaptive anxiety in this population. For instance, does the mother, worried about her child going to the swim club have reason to experience a certain level of anxiety? Or is the family that checks on their child throughout the night experiencing maladaptive anxiety? The research cannot come to these conclusions because of the lack of specificity. Another area of research that could be considered is a quantitative measure of anxiety experienced within this population. It would also be important to identify the most salient triggers of participants' anxiety, through initial factors identified in this

study, as well as additional ones. Last, it would be important to study the impact of anxiety on the condition itself.

Limitations.

Based on the nature of this research, there are some clear limitations to this study. First, the study is limited because it includes only those individuals who have access to computers, indicating possible socioeconomic and cultural biases (Mann & Stewart, 2000). These biases limit the research greatly because of implications related to healthcare access, a knowledge base about LQTS, and other possible variables not yet identified. The second limitation is the limited knowledge about the demographics of the individuals due to confidentiality and the archival nature of the study. This demographic information is pertinent to understanding how culture, gender, and age affect the development of anxiety. The researcher also has limited information about the LQTS diagnosis, such as exact mutation and treatment. This limited information also extends into psychiatric diagnoses, and whether or not the individual has been diagnosed or treated previously. The next limitation is the online nature of the group, which suggests a male gender bias, especially because the user group contains individuals, worldwide (Mann & Stewart, 2000); however, the nature of the group may negate this bias and has yet to be identified. If the overwhelming percentage of participants is male or if the overwhelming percentage is female, this may affect the overall interpretation of how anxiety may develop in the LQTS population. The fourth limitation is the possibility of not gaining access to a substantial number of individuals over a certain age (Mann & Stewart, 2000). The inability to access older and younger individuals affected by LQTS may lead to a bias in how anxiety affects this population. These limitations suggest the possibility of not gaining a complete perspective on the development of anxiety (Mann &

Stewart, 2000). Based on these limitations, the researcher must remain cognizant of possible limitations, including how these could affect results and continue to remain theoretically sound in addition to not assuming causality in the data.

Conclusion.

This research marks a stepping-stone for future research regarding this syndrome. The goal of this study was to start to glean data specifically about what may lead to anxiety responses in this population. Several factors were identified that lead to anxiety related symptomology in participants. Due to the nature of the study, it was difficult to differentiate whether anxiety was maladaptive or adaptive; it became evident that LQTS related experiences created anxiety symptomology in participants. Throughout the data, it became clear that participants endured several different illness experiences that contributed to their anxiety. However, it also became evident that participants had difficulty coping, which further contributed to their anxiety. In the data, it becomes clear that participants attributed their anxiety to a lack of information, lack of clarity and poor support systems, which in turn contributed to poor coping styles. It is difficult to ascertain from the data the exact dynamics that contributed to these attributions, but the need becomes clear for an increased recognition about specific illness experiences and how to intervene effectively. If these psychosocial aspects were addressed in conjunction with medical needs, this encompassing approach could significantly impact a person's quality of life and most likely decrease the risk of the condition.

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Appendix

Diagnostic Criteria for LQTS (Goldenberg & Moss, 2008)

Table 2		Diagnostic Criteria for LQTS	
		Finding	Score
Electrocardiographic†			
Corrected QT interval, ms			
	≥480		3
	460–470		2
	450 (in males)		1
Torsades de pointes‡			
	T-wave alternans		1
	Notched T-wave in 3 leads		1
	Low heart rate for age§		0.5
Clinical history			
Syncope‡			
	With stress		2
	Without stress		1
	Congenital deafness		0.5
Family history 			
	Family members with definite LQTS		1
	Unexplained SCD in immediate family members <30 yrs old		0.5

Scoring: ≤1 point, low probability of long QT syndrome (LQTS); 2 to 3 points, intermediate probability of LQTS; and ≥4 points, high probability of LQTS. †Findings in the absence of medications or disorders known to affect these electrocardiographic findings. The corrected QT interval is calculated by Bazett's formula: $QT/RR^{0.8}$. ‡Torsades de pointes and syncope are mutually exclusive. §Resting heart rate below the second percentile for age. ||The same family member cannot be counted in both categories. Reprinted, with permission, from Schwartz et al. (20).

SCD = sudden cardiac death.

Figure 1. Diagram of initial treatment themes.



Figure 2. Diagram of continued treatment themes.

