A Qualitative Study Examining Factors Related to Treatment Adherence in Persons with Long QT Syndrome

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A QUALITATIVE STUDY EXAMINING FACTORS RELATED TO TREATMENT ADHERENCE IN PERSONS WITH LONG QT SYNDROME

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Submitted in Partial Fulfillment of the Requirements of the Degree of

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DEPARTMENT OF PSYCHOLOGY

Dissertation Approval

This is to certify that the thesis presented to us by Alison B. Steinhauser on the day of Monday May 3, 2010, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

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father. I would have never begun this journey of education with your gentle push and reassurance that I was more than capable. I promise to always work hard and make you proud. “If it was easy, everyone would do it.” I love you.
Abstract

Long QT syndrome (LQTS) is an electrophysiological disorder of the heart affecting as many as 1 in 2,000 persons. LQTS can result in potentially catastrophic and fatal arrhythmias in infants, children, adolescents, and adults. Maintaining adherence to treatment can decrease these negative outcomes and result in favorable quality of life in this population. The present study utilized a qualitative research design to examine and understand nonadherent and adherent behaviors in persons with LQTS. This study was part of a larger study and used a sample from an existing online LQTS user group message board consisting of 780 international members. A total of 68 messages were analyzed, representing 37 global participants. It was found that the overall vagueness in diagnosis and treatment of LQTS and a resulting feeling of loss of control negatively impacted adherent behavior. In addition, it was theorized that several subtypes of compliance may be relevant: precompliance, hypercompliance, and pseudocompliance.
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Chapter 1

Introduction

Statement of the problem.

Long QT syndrome (LQTS) is a rare cardiac disorder caused by a prolonged QT interval corresponding to the repolarization phase of the ventricular action potential (Khan, 2001). Each year, more than 250,000 persons die from sudden cardiac arrest. LQTS is one condition associated with sudden cardiac arrest. LQTS has been of interest to researchers because of the variability in its physiological presentation and the apparent increased number of cases that are being discovered each year. LQTS is a condition with the potential for many psychological effects, but with limited research addressing these psychosocial effects. LQTS is a chronic illness experienced by persons of all ages, with varying degrees of severity and a wide range of implications for human functioning.

As of 2002, more than 1,200 persons with LQTS were identified in the LQTS registry; however, this registry is not a complete compilation of all persons with long QT syndrome, and this number is likely to be significantly higher (Moss & Schwartz, 2003). The number of persons identified with LQTS is increasing as the quality of screening tools improves and the medical field becomes more aware of this potentially deadly condition.

Currently, there is limited psychological research on LQTS. Due to the limited amount of current psychological literature on this disorder, research on other chronic illnesses such as diabetes and asthma can offer information from which researchers can extrapolate similarities in psychological symptomatology. Caution must be exercised when generalizing due to differences in diagnosis and symptomatology.
Compliance is an area of concern in chronic illness management. Persons with LQTS are required to remain on strict medication management programs to minimize cardiac symptoms and prevent death. Medication and treatment noncompliance could result in catastrophic complications for patients with LQTS. Complications include increased symptoms, fainting, and even death. Therefore, research in the area of medication and treatment noncompliance is essential.

**Purpose of the study.**

The purpose of this study was to evaluate medication and treatment noncompliance and nonadherence, particularly in persons with long QT syndrome from patient/caretaker perspective and reports. It evaluated and analyzed common barriers to medication and treatment adherence from parental reports in children and adolescents diagnosed with long QT syndrome. This study examined common threads in the statements and concerns of parents of children and adolescents with LQTS and will provide insight into the causes of medication and treatment noncompliance in this population. The present study was intended to provide insight into some common obstacles that prevent patients from complying fully with their treatment regimens. It is hoped that insight into the possible causes of noncompliance would provide multidisciplinary treatment teams of physicians and psychologists pertinent information that can help them to minimize complications of treatment due to noncompliant behavior.

**Overview of literature review.**

Long QT syndrome is an electrophysiological disorder of the heart that is associated with a lengthening of ventricular repolarization. Persons with LQTS have with a structurally normal heart; however, they experience rhythm disturbances
associated with the lengthened repolarization. Treatment of LQTS includes the use of lifestyle modifications, medications, pacemakers, internal defibrillators, and sometimes surgery on the autonomic nervous system. The subsequent literature review will examine medication and treatment noncompliance in a variety of patients with medical ailments. Varying definitions of noncompliance will be presented and discussed, resulting in a primary definition that will be used throughout this analysis. Compliance and nonadherence research in children and adolescents with diabetes and HIV is reviewed in an effort to examine other chronic illness and compliance. Lastly, the health belief model is examined as a mechanism by which people come to understand their chronic illness and the influence that may have on medication compliance (Janz & Becker, 1984).

**Relevance to cognitive behavioral therapy.**

Medication compliance is critical to the proper care of those with many chronic illnesses, including long QT syndrome. A goal of this research study was to better understand why patients do or do not follow the recommendations made by physicians and other medical professionals to ensure their best treatment. This manuscript aims to provide information that will help psychologists and medical professionals identify obstacles to adherence. It is hoped that understanding barriers to compliance will generalize from the LQTS population to others and provide information to better treat persons who are not compliant with their treatment regimens.
Chapter 2

Literature Review

Advances in medical technology are allowing a growing number of children who are born with special health needs to survive past infancy and into adulthood. Today, babies born with chronic and persistent illnesses have more than a 90% chance of living into adulthood (Blum, 1992; Hagood, Lenker, & Thrasher, 2005). Chronic illnesses require long-term, often life-long management. In recent years, there has been a shift within the medical community towards giving patients a more critical role in their disease management by increasing their role in their treatment regimens. Government agencies, hospital administrators, and physicians are insisting that patients no longer take a passive role in their treatment. Instead, these groups are recommending that patients become fully engaged in decision making, treatment planning, and general management of their chronic illness (Barlow & Ellard, 2004).

With the growing number of persons with chronic illness living into adulthood, there is a new obligation for the medical community. As prognoses improve, there is a need for more long term medical care (Hagood et al., 2005). There is a growing need, and growing concern within the medical profession, on how to best transition pediatric patients with chronic illness through adolescence into the adult medical system. The medical community needs to focus on this transition and consider health care needs, family centered psychoeducation and the developmental issues of adolescence (Hagood et al., 2005).

Society also plays a role in the management of chronic illness through the stigmas and perceptions it places on persons with chronic illness. Often, disability and handicap
are used interchangeably; however, they have drastically different definitions and implications. *Disability* refers to a functional impairment due to physical limitation, while the term *handicap* is controlled by the social context and the limitations society places on these physical limitations (Blum, 1992; Boice, 1998). This misunderstanding may influence how society, peers, and even family members interact with a child who has a debilitating or chronic illness. A child’s perception of his or her own chronic illness can be greatly influenced by societal limits. These limitations can influence how a child or adolescent perceives or understand his or her own disease state and level of functionality. A perceived negative outlook on their disease state, as influenced by a rather uneducated society, can result in reduced hope and a negative outlook on the future, potentially influencing self-care and treatment adherence.

**Chronic illness in children and adolescents.**

Adolescence is a difficult time filled with growth and change. During this developmental stage, children experience biological, psychological, emotional, social, and cognitive growth and change. The presence of a chronic illness is an additional stressor during this already tumultuous time. As more and more children live past childhood and into adolescence and even adulthood with chronic illnesses, there has been increasing interest in the psychological ramifications that chronic illness may cause in adolescent/adult patients and their families. As this manuscript focuses on children and adolescents, the remainder of this review will focus on psychological effects found in these populations.

Blum (1991) examined the psychological effects that may be experienced by children and adolescents living with chronic illness. Blum categorized areas of concern
that are specific to adolescents and influence their ability and method of coping with chronic illnesses.

**Social-emotional development.**

Blum (1992) points out several factors that affect the degree of impact the diagnosis of a chronic illness has on the social and emotional development of a child or adolescent. These factors include level of incapacitation or impairment, prognosis, illness symptomatology, and cost (Blum, 1992). Children and adolescents are greatly impacted by the degree to which their chronic illness prevents them from being mobile. Interestingly, mild physical limitations are often as difficult or more difficult to cope with than more severe immobility. Persons with invisible rather than outward disabilities often have more emotional problems. There is the belief on the part of the patients that their illness limitations may be discredited because they is not visible. For illnesses that are not outwardly obvious, there are concerns about revealing the chronic illness. There is evidence to suggest that children with chronic illnesses who appear healthy have more adjustment problems because of their perceived need to always justify and explain themselves (Boice, 1998). As such, persons with LQTS may have difficulty coping with their condition because it is not a visible, predictable, or continuous outward disability.

Lack of clarity about prognosis also negatively affects children and adolescents. Those patients with uncertain prognoses often have more difficulty adjusting and coping than those patients with prognoses of death. Patients with LQTS constantly negotiate an uncertainty about their prognosis because a sudden cardiac arrest could occur at anytime. Furthermore, patients with LQTS may, or may not, experience symptoms periodically throughout their lives. Patients who suffer from chronic illness that remit and return find
their disease more burdensome than those patients whose disease states are truly chronically unrelenting (Blum, 1992; Boice, 1998).

Children and adolescents with chronic illnesses feel different from their peers (Felton & Revenson, 1987). These children are often socially and developmentally delayed because they have not had equal exposure to social experiences as compared to healthy children (Blum, 1992). Feeling ill, hospitalizations, doctors’ appointments, and even parental restrictions influence the amount of socialization children/adolescents with chronic illnesses are exposed to. Children and adolescents with LQTS may have few hospitalizations; however, they may experience significant restrictions and numerous limitations in their daily lives. The period of adolescence and the occurrence of puberty make this stage a vulnerable time. It is during this time that being different from one’s peers, especially in limitations, is especially difficult (Blum, 1992; Boice, 1998). The perception of being so different from one’s peers may result in negative cognitions and emotions. Normal issues experienced during adolescence may be magnified or enhanced in children and adolescents with chronic illnesses.

**Development and growth.**

The development from being a dependent child to an independent adult requires that an adolescent making this journey take responsibility for their actions, weigh their needs against the needs of others, and develop supportive relationships (Blum, 1992). There are physical, psychological, and cognitive changes that affect the way in which children and adolescents cope with their chronic illness. From a physical standpoint, chronic illness can affect growth and development. Growth and puberty may be delayed or affected by the presence of illness during development (Boice, 1998).
Teenagers also begin to require and demand autonomy. Autonomy in adolescents is an earned commodity and is dependent on parents feeling comfortable in letting their children grow. Chronic illness in children and adolescents require that parents feel comfortable letting their child take a chance and believe their child will be safe and able to care for themselves (Boice, 1998). Allowing a chronically ill child to explore his or her autonomy requires that parents recognize the child’s physical condition while not viewing them as handicapped (Blum, 1992). This can impede a child from feeling independent and autonomous when they strive for that developmental goal (Orr, Weller, Satterwhite, & Pless, 1984). There are several times in LQTS patients’ development where a struggle for independence may take place, for example, allowing a child to go to friends’ homes, or going on an overnight visit and being responsible to take his or her medications, or trusting a child or adolescent will keep himself or herself hydrated during a warm day at school or while out with friends. Parents have the difficult job of navigating the desire and need for their child to develop and reach independence while calculating the level of autonomy their child can handle at any given developmental stage.

Adolescence is a developmental stage marked by great egocentrism. Adolescents at this phase in their lives tend to believe that they are of particular interest to other people. This phenomenon is known as imaginary audience and personal fable. Having this belief, the adolescent believes that everyone is looking at and concerned with them. Adolescents with chronic illness may be overly sensitive to this feeling of being watched because they are often the center of attention at home and the center of attention while receiving medical attention. In addition, there is also a belief held by many teenagers that
they are completely unique in their experiences. Children with chronic illnesses may feel even more unique in their experience because they are a minority (Orr, Weller, Satterwhite, & Pless, 1984).

**Family issues.**

As previously stated, chronic illness impacts not only the life of the patient, but also the entire family unit. The topic of family issues can be represented from the perspectives of both the teen and parents. From a parental perspective, the knowledge that their child will become an adult with specific limitations raises some very poignant areas of inquiry for parents. As the child enters puberty, questions about adulthood and independence begin to emerge. These questions include genetic risks, specifically the likelihood that the child is at risk for passing along genetic information that will result in another chronically ill child. Life expectancy is also an area of concern. Parents and physicians may be unsure about the long-term survival of the child. Academic and vocational potentials are also of concern to parents as they begin to conceive of their children needing to become independent of them (Blum, 1992). Families with chronically ill children tend to be more socially isolated and therefore do not have external social supports. The lack of an extended support system restricts coping and can have a negative outcome on the family members, who have to rely entirely on each other.

The chronically ill child also has his or her own concerns and issues surrounding their family situation as it relates to their illness. As mentioned, adolescence is a time of emerging autonomy and certain family situations can impede the desire to become more independent. Often, the typical family conflicts that arise when a child is going through puberty and entering adulthood are magnified and enhanced in families with children
with chronic illnesses. As mentioned, families of chronically ill children tend to be isolated. During adolescence, the growing teen desires more time away from the family in an effort to explore his or her individuality. Parents who are enmeshed with their children and lack support from outside of their home may become overly burdensome to the young adult who is trying to develop a sense of self.

Overprotection is a common problem in families with a chronically ill child. Overprotection comes in many forms, including parents providing too much assistance for everyday tasks, being overvigilant, issuing constant reminders about self-care, and being overly restrictive. While some of the restrictions and hypervigilance parents provide is appropriate for children and adolescents with LQTS, an adolescent may react with anger, rebellion, or resentment. Adolescents perceive overprotection as evidence of their being incompetent (Blum, 1992; Boice, 1998).

Coping with chronic illness: Children versus adults.

The process by which patients adapt to the diagnosis and resulting management of a chronic illness is of particular interest in both the medical and psychological arenas. The ability to effectively adapt to and cope with chronic illness is imperative due to the permanence of the disease and the resulting life-long management. The ability to effectively cope in childhood can have a significant impact on how patients cope in adulthood. Put simply, positive coping skills in childhood can result in positive coping abilities in adulthood (Bauman, Drotar, Leventhal, Perrin, & Pless, 1997; Pless, Power & Peckman, 1993).

The long-term consequences of chronic illness make the process of adjusting to chronic illness a difficult but necessary phenomenon. There is a widely held belief
among clinicians that children with chronic illnesses are at higher risk of having emotional, social and behavioral complications (Bauman et al., 1997; Pless et al., 1993). The vulnerability to maladjustment later in life is concerning. Schmidt, Peterson, and Bullinger (2002) propose that effective coping in childhood can significantly impact adjustment and coping in adulthood. Children with chronic illnesses who learn to effectively adapt to and cope with a diagnosis in childhood are less likely to present with psychosocial, emotional, and behavioral problems later in life (Schmidt et al., 2002). Treatment nonadherence may be the result of poor coping with and nonacceptance of the medical necessities at hand.

**Coping: Developmental perspective.**

Coping abilities differ for adults and children. This difference is primarily because of their developmental stage and capacity. The ability to cope is dependent on cognitive development, emotional development, and personality development. In contrast to adults and adolescents, children tend to be very concrete in their ability to cope because they are limited to behavioral strategies as a method of coping. With age, however, children have less concrete thinking and begin to develop and utilize their growing cognitive abilities. Adolescents develop cognitive and emotional coping skills as they get older. Because of this, adolescent coping tends become more fluid and flexible. This development follows Piaget’s stages, following a child from concrete to formal operations (Schmidt et al., 2003).

**Piaget’s developmental theory.**

At the heart of Piaget’s developmental theories are structure and function. According to Piaget, structure and function are two types of heritable realities which
influence the development of reason. Piaget describes structure as organs or qualities that are the instruments for intellectual adaptation (Piaget, 1952). The structure refers to the particulars of an event or behaviors performed by the subject. It would include such behaviors as looking at medication, reaching for water, taking the medication, and following medical advice. Each of these tasks is related, which gives it structure.

According to Piaget, function refers to how humans receive the information through our structure. He adds that function refers to how we inherently, or naturally, interact with our environment using all of our biological systems (Piaget, 1952). While functions are stable over the lifespan, structures change as the individual develops. Without this flexibility, development would not be possible.

Piaget articulated four stages of development that begin at birth and conclude around the age of 15. These stages include the sensorimotor, preoperational, concrete operational, and formal operational stages. The sensorimotor period of development begins at birth and typically continues until approximately 2 years of age. It is a period of rapid growth and development in which the child develops many new cognitive structures, without the use of language (Phillips, 1969; Zigler & Gilman, 1998). Without the ability to use language as a method to learn and understand, the infant relies on sensory systems and the ability to use his or her limited motor skills to manipulate objects to learn about the environment. At this stage of development, children are unable to take the perspective of another person. Piaget refers to this as egocentrism. This egocentric understanding of the world makes it difficult for children to conceptualize how objects or people can exist outside of them. At this point, if an object is not in their view it simply does not exist. For example, if you hide a toy from a 1-year-old child, he or she will not
search for it (Zigler & Gilman, 1998). This ability changes once the child has an understanding that objects and people can and do exist outside of their perceptual field. This understanding is known as object permanence, that is, objects continue to exist even when you cannot actively see them and is acquired at the end of the sensorimotor period.

During the sensorimotor stage of development, children would have essentially no ability to care for themselves in any medical capacity. Not having the understanding of how outside organisms such as medicine can impact them limits their ability to perceive this as a necessity in their lives.

The second stage of development is referred to as the preoperational stage of development and extends from approximately the second year until the seventh year. During this stage, children are able to mentally represent symbols and manipulate these symbols to represent a difference in their environment in limitless ways (Phillips, 1969). This ability allows them to truly begin engaging in imaginative play. During this time, children are extremely egocentric and unable to see another person’s perspective. They have a very difficult time mentally representing a reversal of an action or anything more than a single dimension.

A child in Piaget’s preoperational stage of development could begin to utilize symbols and imaginative play to understand the body’s medical requirements. A 3-year-old child with long QT syndrome in the preoperational stage of development may use imaginative play to give his/her teddy bear medicine or pacemaker checks for example. The child may also tell his or her toys what they may or may not do in an effort to play out their treatment adherence requirements. Children in this stage of development can
begin to integrate the importance of compliance; however, it still requires the assistance of an adult to monitor and provide a structured compliance environment.

The concrete operational stage of Piaget’s theory begins at approximately 7 years. At this stage of development, children are now able to represent more wholly the aspects of their physical environment. Children are able to manipulate or transform things mentally that they previously would have had to physically manipulate. They are able to mentally represent physical things; however, they are still unable to think in complete abstractions (Zigler & Gilman, 1998). Children with LQTS in the concrete operations stage of development are able to conceptualize the importance of the treatment regimen. For example, they begin to understand the need for all adherence measures (taking medications, avoiding caffeine, avoiding certain activities, keeping doctors’ appointments); however, they are not able to conceptualize how these adherent behaviors work in conjunction to prevent symptoms.

The final stage in Piagetian development is formal operations. Formal operations typically arise after the 12th year; however, not everyone enters this final developmental stage (Zigler & Gilman, 1998). The formal operational stage of development is marked by the ability to use the hypothetical. This stage of development allows children to think about the possibilities and options that they have. They are no longer locked into concrete, tangible thinking. This is also the stage when older children become capable of completing more complex problems and learning the experimental process. At this entrance into adolescence, the individual begins to use the ability to form hypotheses and deduce alternatives. This advanced level of thought and logic is advantageous in the ability to problem solve and interact in the environment in a more productive and
efficient way. Adolescents with LQTS who enter the formal operational stage of development are fully capable of understanding the consequences of noncompliant behavior. Furthermore, adolescents in the formal operational stage of development also become cognizant of their options and may begin to rebel or seek more independence from their caretakers.

**Technical and adaptive competence in coping.**

To further understand and explain the development of adaptive coping, Schmidt et al. (2003), using an earlier model by Fournet et al. (1998), distinguish between technical competence and adaptive competence in the development of adaptive coping. Technical competence refers to a person’s cognitive, emotional, and personality traits and the flexibility that a child has to work within them. Adaptive competence is the way in which a child responds to situational stress. It is the combination of these two competences that dictate the degree to which a person is able to effectively cope (Schmidt et al., 2003).

Just as children become less concrete and more fluid as they develop into adolescence and adulthood, children also shift between technical competency and adaptive competency. As younger children tend to be more behaviorally oriented in coping strategies, they also rely on technical competencies to alleviate their stress. Their concrete nature does not allow young children to clearly navigate a problem or explore their resources. As children grow and begin to develop, their adaptive competency increases. This allows them to better distinguish between situational demands, their desires, and perceived outcomes. As a child or adolescent develops and begins to use adaptive competencies to cope, he or she begins to understand the situational demands of
the illness (Schmidt et al., 2003). In a patient with LQTS, such situational demands may include maintaining a medicine regimen, decreasing or ceasing some physical activities, and complying with various other restrictions. In their ability to use adaptive competencies, children or adolescents with LQTS begin to recognize the perceived outcomes of maintaining compliance versus noncompliance. Measuring perceived outcomes is a function of the health belief model, which will be further examined later in this literature review.

**Positive effects of early coping.**

A study conducted by Hampel et al. (2005) suggests that successfully coping with a chronic illness early in life may lead to better overall coping abilities later in life. Hampel and coworkers examined 156 patients between the ages of 8 and 13 with chronic illnesses such as asthma ($n = 47$), atopic dermatitis ($n = 52$), or cancers ($n = 57$) and a group of healthy controls ($n = 158$). The researchers used the German Coping Questionnaire for Children and Adolescents, which rates nine coping strategies on a Likert scale. The results indicated that children with chronic illness engaged in fewer avoidance behaviors than their healthy peers. Results also indicated that while their coping abilities may be more efficient than those of the healthy controls, adolescents between the ages of 13 and 15 are at increased risk of developing psychological effects because of lack of their coping abilities, possibly due to increased social stressors. Additionally, results indicated that adolescent females in particular are at increased risk for poor coping (Hampel et al., 2005).
The effect of coping on the family unit.

Chronic illness in a child greatly impacts the lives of each family member. The child is aware that his or her illness impacts not only their own life, but also the lives of their parents and siblings. Children with chronic illnesses tend to be more dependent on their family unit and more submissive in their relationships. Adolescents are developmentally at a stage at which they are striving to become more independent. Chronically ill adolescents tend to be more dependent than their healthy peers. Speaking more globally, chronically ill children and adolescents are more dependent in many facets of their lives. This population is dependent on their parents, physicians, medical technology, and medications. Children and adolescents with LQTS are dependent upon their parents, their medical team (often several members), oral medications, and an automatic external defibrillator (AED), implanted cardioverter defibrillator (ICD), or pacemaker.

Further, family members often reinforce the chronically ill child’s dependence by decreasing the task demands (when demands are appropriate) that they place on the child. For example, a 6-year-old with LQT3 may be excused from taking out the garbage as a household chore; however, this chore is appropriate for the child and would likely be acceptable for someone with LQTS. Expecting less from an ill child, and requiring that they do less as a family member, negatively impacts their ability to gain independence (Schmidt et al., 2003).

Family coping styles.

As the requirements of day-to-day care shift from being the responsibility of health care providers to the responsibility of the patient and the patient’s family, the
requirements of persons in caretaking roles are affected. In the case of children and adolescents with chronic illnesses, the responsibility of management and treatment involves the entire family. The chronic illness becomes a “family affair,” and the parents and even siblings play important roles in the care of their loved one (Barlow & Ellard, 2004). When chronic illness “invades” a family, there is an intense psychosocial impact on the entire family. The stress surrounding the presence of a chronic illness and the resulting management requirements can be very taxing on all family members. An impact is experienced by the family emotionally, physically, and socially. Coping styles influence how deeply the stress of chronic illnesses such as LQTS influence each individual family member and the family unit in its entirety (Feigin, Barnetz, & Davidson-Arad, 2008).

In families affected by LQTS, one or more family members may have the disease. If both a parent and child have LQTS, the parent is in the dual role of being both a patient and a caregiver. It is possible that the healthy spouse would feel more responsibility, as it would be his or her role to be caretaker to both the spouse and the child with LQTS. There are many family dynamics that can be influenced by having multiple family members with LQTS. Issues may arise concerning parenting roles and differing sibling roles, as LQTS may impede daily functionality. Thus, the division of labor in a family would be unbalanced, and this could lead to hostility, resentment, or general discomfort within the family dynamic. In addition, consideration of the impact of having multiple members with LQTS or a child or sibling who does not have LQTS yields other possible family dynamics.
**Family management styles.**

As previously stated, children with chronic illness require the support, both emotional and physical, of their family. Knafl, Breitmayer, Gallo, and Zoeller (1996) conducted a qualitative study that examined family response styles to childhood chronic illness. Knafl et al. used a sample of 63 families with school-age children (7 to 14 years old) with chronic illness. Families were interviewed with open-ended questions on two separate occasions. Researchers identified five unique family management styles: thriving, accommodating, enduring, struggling, and floundering. Family management styles (FMS) can be understood as the way in which a family configures itself, based on individual family members’ perceptions of their situation and the way in which they manage their chronic illness (Knafl et al., 1996). The family management style that is adopted within the family unit determines how the family copes with the diagnosis and determines individual self-views, management approaches, compliance, parental relationships, and future outlook. Examining the way in which family members experience the subjective meaning of the situation, as well as the management behaviors family members adopted, determines the success of management and affects the interaction within the family (Knafl et al., 1996).

Compliance and treatment adherence can be influenced by family management style. Treatment adherence may be influenced by a family’s confidence in their ability to effectively care for their loved one. The perspective a family maintains about a chronic illness, such as long QT syndrome, and to what degree they make LQTS central in their lives can influence treatment adherence. Keeping a positive perspective and an optimistic outlook on the longevity of their loved one can increase the likelihood that
they will follow the physicians’ directions and be adherent to treatment. Being optimistic that treatment is useful and beneficial, in other words the perceived benefit of the treatment modalities is highly regarded, increases compliance (Janz & Becker, 1984). Furthermore, maintaining normalcy and routine in the family will behaviorally increase compliance, while a negative perspective of the illness and treatment modalities may interfere with compliance because it may feel too burdensome or overwhelming, as well as out of their immediate control.

**Thriving family management style.**

Thriving families are defined as those who are confident in their ability to manage the child’s chronic illness. All family members take a proactive stance in illness management and incorporate medical regimens into daily routine. In families with LQTS, this would be evidenced by having the entire family involved in daily treatments, such as medications. Equal care would be taken by all family members that proper physical requirements and restrictions are met. An example of such behaviors could be demonstrated between siblings. A sibling relationship in which one sibling has LQTS could involve the other child by having the healthy child take vitamins as her/her brother/sister takes their daily medications or making a family decision to have a caffeine-free home. Families identified as having a thriving FMS perceive their ill child as normal, or equal to their healthier peers and family members. This concept of normalcy is an overarching concept in this management style. A thriving family with LQTS would not perceive the illness or the family member who has LQTS as negatively impacting their family because they do not place the illness at the center of the family’s life. In a thriving family system, LQTS would become part of the life, not the center of
it. Because they do not focus on the illness or view it as a negative thread that runs through the family, thriving families maintain a bright future for their family’s happiness and for the happiness of their child (Knafl et al., 1996).

**Accommodative family management style.**

Like the thriving family management style, those families who adopt the accommodative family management style also focus on keeping normalcy within the family. However, these families have a more negative perspective towards chronic illness management. Some of the parents in this group viewed their child’s illness and the influence that it had on their family as tragic. The trajectory of the illness was perceived as negative, and the ill children expressed more worry about their long-term quality of life. Specifically, these families viewed the child as a tragic figure whose life will be forever compromised, unfulfilling, and ominous. The chronically ill children in the accommodative families viewed themselves as less healthy than their peers and other family members. While mothers of accommodative family management styles viewed themselves as competent caretakers, fathers in this FMS tended to be less involved and viewed themselves as uninvolved in illness management. Accommodative parents mutually respected their spouses’ role within the family structure and expressed gratitude for their respective roles (Knafl et al., 1996).

A family with LQTS who adopts an accommodative FMS may find itself struggling with the prognosis of LQTS. An accommodative family system may interpret the daily regimen of medications, restrictions, and future invasive treatments (if necessary) in an ominous light. This approach to the future of the patient leaves the family member with LQTS feeling sicker and more limited than they are. Family members may not evenly
divide the responsibility of care (as seen in the thriving FMS), resulting in a single caretaker feeling overwhelmed by responsibility.

**Enduring family management style.**

The enduring family management style is dominated by a negative view of the family’s situation, as well as a negative feeling towards the effort required to care for a chronically ill child. Similar to the accommodating FMS, these families view their child as a tragic figure; however, families with an enduring FMS take this belief one step further and perceive their child as tragic and having a irreparably compromised future (Knafl et al., 1996). A family coping with LQTS with an enduring FMS may view their child or loved one as being doomed or having an excessively negative future. They may perceive their loved one as having a future that cannot bring happiness or a good quality of life. In this perspective, LQTS has robbed, in a sense, their loved one of any semblance of a normal, happy, and productive life.

Parents with enduring FMS tend to place more restrictions on their chronically ill child, in an effort to protect them from possible harm. There is a strong emphasis on setting limits, restrictions, and teaching the child to accept such restriction in their lives. A child with LQTS may be restricted from spending time away from home with their peers and be restricted from engaging in any “normal” childhood activities for fear of a tragic consequence. Parents in the enduring FMS perceive the work required to care for their chronically ill child as very effortful. There is a sense that the illness is a burden on the family, and there is an inability to conceive of ways to make illness management less burdensome.

**Struggling family management family style.**
The struggling FMS is characterized mainly by parental conflicts. These families experience considerable conflict within the family, typically centered on the fundamental differences in how each parent views the family’s situation and what they expect of their partner. There is often blame placed within the parental relationship because the division of labor is unclear. Mothers in this group reported receiving little support from their husbands, claiming that they are insufficiently involved and unappreciative. The fathers’ uninvolve ment increased the perceived burden felt by mothers. Furthermore, mothers typically viewed their child’s future as grim and experienced fear of the future (Knafl et al., 1996).

A struggling FMS dealing with LQTS would have a great deal of conflict. The uneven distribution in the caretaking role and the resulting resentment or anger may result in an uncomfortable family dynamic. The result of this uneven division of labor, and the resulting hostility, would inevitably affect LQTS patients by adding additional stress to their lives and creating the feeling that they are a burden and source of conflict to their other family members. A struggling FMS may lose focus on the importance of treatment compliance because they are overwhelmed by the stress of the chronic illness. The primary conflict in families with the struggling FMS is the lack of agreement about how the family situation can be improved. While neither parent would be content with the current state of their family, a compromise would not be found. The struggling family perceives the child’s chronic illness as a central focus of the family (Knafl et al., 1996).

*Floundering family management style.*
The floundering FMS is defined primarily by confusion and negativity. Parents of floundering families perceive the child as a tragic figure and have an intolerant stance towards the restrictions placed on both the child and the family. A floundering family may deny the need for strict illness management and become nonadherent to treatment recommendations. Within this family dynamic, there is a theme of uncertainty. Primarily, parents are unclear on how to best care for the child or the family. Families with a floundering FMS had a difficult time managing the illness, and therefore, proper management and adherence was not maintained.

Parents in these families perceive the management of the child’s illness as a burden and feel inadequate in the ability to effectively care for the child. Their lack of confidence resulted in problems only being addressed when they became serious. A family with LQTS with a floundering FMS could have catastrophic results. LQTS requires that proper preventative measures be taken (medications, device checks, regular doctors’ visits), and the floundering family may not be able to provide adequate preventative care. Their negative view of the family situation infused into their perception of the child’s future results in the illness taking a central role in the family’s dynamic (Knafl et al., 1996).

These five family management styles that Knafl et al. identified can assist health care workers to better understand the dynamics of families with chronically ill children. These categories can help health care workers to make sense of the illness from the viewpoint of the family, as well as of the meaning the illness has within the family. The way in which a family understands and adapts to a diagnosis influences subsequent
family dynamics, parental communication, family routine, and treatment compliance, as well as the way in which families frame chronic illness within the child’s life.

**Family interventions.**

The importance of the role of the family unit in health and disease management is evident. Campbell (2003) outlined and reviewed evidence that families impact both health and treatment and offered a typology of different interventions than can be used within families. Campbell indicates that there are several findings about chronic illness and the family dynamic. First, families have a large influence on individual members’ health. Social support within a family has been found to be health promoting. Persons with LQTS would benefit from having a family structure such as the thriving family management style because of the physical and emotional support that is provided and the team approach the family takes towards management.

The second relevant conclusion found by Campbell indicated that emotional support provided by family members is the most important type of support that one can receive. Other methods of support, such as providing the logistical arrangements, caretaking (dispensing medications, preparing appropriate meals), or providing information were not as influential as providing emotional support for the person with a chronic illness (Berkman, 1995; Campbell, 2003). From an LQTS perspective, the family support provided to the family member(s) with the condition is the most influential support provided. It imparts a sense of belonging and feeling loved and cared about.

Lastly, hostile or negative family dynamics such as the floundering and struggling family management styles have a stronger influence on health and disease management than do positive family dynamics. A negative or poorly adjusting family can do more
harm than a positive and supportive family can help a person with chronic illness. Negative environments induce stress which could increase physical symptoms (Berkman, 1995; Campbell, 2003). A person with LQT1 with living in a non-supportive hostile home may feel stress which elevates his or her heart rate to dangerous levels. Or, a LQT2 patient may become emotionally stressed or be startled when family members argue.

It is critical that the medical team articulate to the family the importance of a supportive, caring, and loving environment. The impact that a family has on the patient is immeasurable. A supportive family can help a child, adolescent, or adult with LQTS adhere to the treatment recommendations and can control family distress and distress experienced by the patient. Long QT syndrome is a complex diagnosis that influences and impacts all parts of individuals’ and families’ lives.

**Long QT syndrome.**

Long QT syndrome, often referred to as LQT and LQTS, is estimated to affect 1 in 2,000 people (Schwartz et al., 2009). As one of the leading causes of sudden death in children and adolescents, LQTS has been receiving increasingly more attention and research since the first family with LQTS was discovered in 1957 (Chiang, 2004; Goldberg & Moss, 2008; Kass & Moss, 2003). Long QT syndrome is an electrophysiological disorder of the heart that is associated with lengthening of ventricular repolarization of the heart. LQTS patients present with a structurally normal heart; however, the patient is susceptible to an arrhythmogenic complication associated with QT interval prolongation and electrical instability in the heart. (Goldenberg & Moss, 2008; Khan, 2001a, 2001b; Napolitano, Bloise & Priori, 2006).
Long QT syndrome was first identified in 1957 by Jervell and Lange-Nielsen. They studied a family that consisted of six children, four of whom were deaf and experienced syncope. Three of these symptomatic children died suddenly while playing at 4, 5, and 9 years of age. Electrocardiograms (ECGs) of the other three children displayed a prolonged QT interval (Chiang, 2004; Goldberg & Moss, 2008; Kass & Moss, 2003). Later, this disorder would come to be known as Jervell and Lange-Nielsen syndrome and is also known as LQT1. This is only one expression of the many types of LQTS that have been identified in patients (Chiang, 2004; Sovari et al., 2008).

**Acquired versus congenital long QT syndrome**

Long QT syndrome can be identified as having a congenital or acquired origin. Congenital LQTS is a heritable disease associated with gene mutations that affect the efficiency of ion channels (Arnestad, . Acquired long QT is typically caused by medications, including antibiotics, antidepressants, antifungals, diuretics, and psychotropics, in genetically susceptible individuals. It can also be precipitated by cocaine abuse, poison ingestion, strokes, fasting or a variety of other causes (Khan, 2001a; Mayo Clinic, 2009). The majority of research focuses on congenital long QT, and for the remainder of this literature review, long QT syndrome will be understood to mean the congenital type.

**Long QT syndrome explained.**

As previously mentioned, LQTS is a disorder affecting the repolarization phase of the action potential related to abnormalities in ion channels (Goldberg & Moss, 2008; Kahn, 2001a; Kass & Moss, 2003). The period of time after ventricular excitation and during ventricular recharging is also known as repolarization. This period of
repolarization, when measured in a normal heart and normalized for variable heart rates, is approximately 450 milliseconds (ms) long. This rather long period provides time for the heart to adequately recharge and fill during diastole prior to systolic contraction and affords the heart protection against premature excitation (Kass & Moss, 2003). The abnormal QT interval is demonstrated as prolonged past these normal values (450 milliseconds) on an ECG. The principal causes for the abnormal QT prolongation are a decrease or delay in cardiac ion channels that affect the repolarizing potassium current or abnormalities in calcium and sodium channels and currents (Goldenberg & Moss, 2008). The abnormality in the potassium current is due to mutations in subunits of ion channels (either alpha or beta) that affect the cardiac potassium currents. Defects in the sodium channels found in persons with LQTS (primarily LQT3) result in a small number of sodium channels remaining open and conductive, rather than becoming nonconductive during the repolarization (Goldenberg & Moss, 2008; Kass & Moss, 2003).

**Genetics and long QT syndrome.**

Long QT syndrome typology is defined by the mutation that is found. There have been hundreds of genetic mutations and more than 12 variations of LQTS identified. These variations are, LQT 1 to 6, LQT 7 (Andersen syndrome), LQT 8 (Timothy syndrome), LQT9 to 12, JLN1, and JLN2. However, even with the growing number of mutations and variations being identified, there are three most common mutations accounting for the vast majority of cases (Sovari et al., 2008). These are LQT1, LQT2 and LQT3 (Goldenberg & Moss, 2008). Phenotypical presentation of LQTS is based on the presence of a number of symptoms and ECG manifestations. The two earliest descriptions of long QT syndrome were Romano-Ward syndrome and Jervell and Lange-
Nielsen syndrome (Kapetanopoulos, Kluger, Maron, & Thompson, 2006). Romano-Ward syndrome is an autosomal-dominant disorder that does not cause deafness. Jervell and Lange-Nielsen syndrome is an autosomal-recessive trait that is a less common occurrence, but results in a more severe expression of LQTS and presents with congenital deafness (Goldenberg & Moss, 2008; Kapetanopoulos et al., 2006). As medicine advances and more genes associated with LQTS are identified, more types and variations of the syndrome are likely to be discovered. Each respective type of LQTS is sensitive to different environmental and internal factors. Therefore, the triggers for each type vary.

**Long QT1.**

LQT1 is found in 42% to 55% of persons with LQTS (Kapetanopoulos et al., 2006). LQT1 is identified on the ECG as broad based T-waves. The LQT1 mutation affects the potassium channels, delaying repolarization, thus leaving the patient more vulnerable to an early depolarization. Persons with LQT1 have higher frequencies of cardiac events when engaging in activity (Collins & Van Hare, 2006; Goldenberg & Moss, 2008; Kapetanopoulos et al., 2006; Wehrens et al., 2002). In patients with long QT1, the primary trigger is exercise. More than 62% of cases of syncope or sudden cardiac death are the result of exercising, with 26% being attributed to emotion and only 3% occurring during sleep or rest. For persons with LQT1, swimming is a common risk factor and often a trigger of a cardiac event. In fact, nearly one third of all cardiac events take place while LQT1 patients are swimming (Kapetanopoulos et al., 2006).

**Long QT2.**

Long QT2 is identified as a low amplitude and notched T-waves and is found in approximately 35% to 45% of persons with LQTS (Collins & Van Hare, 2006;
Kapetanopoulos et al., 2006). The LQT2 mutation affects the potassium alpha-channel, resulting in prolonged repolarization of the potassium channel. The phenotypic expression of LQT2 is associated with being most at risk or developing symptoms when experiencing emotional stress, and when startled by a loud unexpected noise. Approximately 43% of LQT2 patients experience syncope or sudden cardiac death when surprised or emotionally stressed and only 13% when engaging in exercise (Collins & Van Hare, 2006; Goldenberg & Moss, 2008; Kapetanopoulos et al., 2006; Wehrens et al., 2002).

**Long QT3.**

Long QT3 is identified as a late peaking T-wave after a prolonged ST segment. LQT3 is found in approximately 8% to 10% of persons with LQTS (Collins & Van Hare, 2006; Kapetanopoulos et al., 2006). The LQT3 mutation affects sodium channels responsible for opening with the initiation of an action potential, resulting in depolarization. In persons with LQT3 mutation, the sodium channel fails to inactivate and results in a continued inward flow of sodium. Persons with LQT3 predominantly experience cardiac events during sleep or rest. Statistically, LQT3 patients experience 39% of their cardiac events during rest and 13% while engaging in exercise. While at decreased risk of experiencing cardiac events, patients with LQT3 are more likely to experience cardiac events that are fatal (Collins & VanHare, 2006; Goldenberg & Moss, 2008; Kapetanopoulos et al., 2006; Wehrens et al., 2002).

**Long QT syndrome: Making a diagnosis.**

Diagnosis of long QT syndrome is primarily based on ECG results, family history, clinical assessment, and genetic testing. The two most critical diagnostic
elements are the prolongation of the QT interval and stress-induced syncope. Because some form were easier to identify than others, and standardization was needed, a quantitative approach to diagnosing LQTS was developed. The criteria are divided into three major categories (electrocardiographic findings, clinical history and family history), and each category has subcategories (for example, QT interval length, T-wave morphology, syncope, etc.). The presence of any diagnostic feature has a related point value ranging from 0.5 to 3.0 points. Persons scoring less than 1 point have a low probability of having LQTS, persons with 2 to 3 points have an intermediate probability of having LQTS, and finally those with more than 4 points have a high probably of having LQTS (Chiang, 2004; Crotti, Celano, Dagradi & Schwartz, 2008; Kass & Moss, 2003; Khan, 2002; Naik, 2007; Wehrens et al., 2002).

As previously mentioned, the length of the QT interval is a primary diagnostic element in LQTS. The QT interval measure used to determine if a patient has LQTS should come from a 3- to 5-wave cycle and should always be measured from the beginning of the QRS to the end of the T-wave (Chiang, 2004; Goldenberg & Moss, 2008). The length of the QT interval is most commonly measured using the Bazett Correction Formula, calculated as \( QTc = QT \times \frac{RR}{2} \). QT intervals longer than 440 ms are considered by some to be prolonged. However, there is variation in the values considered prolonged, based on both age and gender. Bazett-Corrected QTc values that are considered prolonged are over 460 ms in children under 15, greater than 450 ms in adult males, and greater than 470 ms in adult females. When conducting an ECG assessment and measuring QT values, the longest value should be used. It is important to consider that the Bazett formula has clinical limitations for heart rates that are unusually
fast or slow. Lastly, it is recommended that several ECGs be obtained while gathering information for diagnosis because QTc values do vary within patients over time (Chiang, 2004; Crotti et al., 2008; Kass & Moss, 2003; Khan, 2002; Naik, 2007; Wehrens et al., 2002).

A second principal diagnostic feature commonly found in LQTS diagnosis (though not necessary) is a history of unexplained syncope, especially during exercise. Syncope is the sudden loss of consciousness in a person. In LQTS, the majority of first events of syncope occur during childhood (Chiang, 2004; Kass & Moss, 2003; Wehrens et al., 2002). In symptomatic patients, approximately 50% experience their first cardiac event (e.g., syncope) by the age of 15. Often, an unexplained syncopal event is what triggers a family to explore a medical causality. Syncopal events are commonly caused by torsades de pointes, a rapid and malignant ventricular arrhythmia. When this torsades de pointes heart rhythm continues without termination, it degenerates to ventricular fibrillation and sudden death occurs (Wehrens et al., 2002). Torsade de pointes often self-terminate, resulting in a fainting experience.

The probability of having syncopal events is dependent on environmental factors and the patient’s specific type of LQTS. Environmental factors may be adrenergic stimuli. Examples of adrenergic stimulation include exercise and emotional or stress-related events. The genotype or specific type of LQTS, in combination with adrenergic stimulation, can result in cardiac events such as syncope or sudden cardiac death (SCD) (Kapetanopoulos et al., 2005).

Persons may present with the complaint of “fluttering” or palpitation experiences if the ventricular arrhythmia or TdP is short lived. However, because torsades de pointes
Arrhythmia often results in syncope or a cardiac arrest, these patients often present to a medical facility with syncope, seizures, or cardiac arrest (Chiang, 2004; Kapetanopoulos et al., 2005). Often, symptoms are increased in females during the time of their menstrual period as well as in the postpartum period, particularly in the LQT2 subtype.

**Sudden cardiac death.**

Sudden cardiac death (SCD) is the result of fatal arrhythmias (Meyer, Mehdird, Salem, Kulikowska, & Kulikowksi, 2003). The length of the QT interval is a significant risk factor in cases of SCD. Patients with QTc intervals that are longer than 500 ms are two to three times more likely to experience SCD. Other risk factors that increase the likelihood of such an event are early morning hours, history of prior cardiac arrests (revived), repeated syncope, congenital deafness, longer QTc intervals, being female, and a history of sudden death in the family (Khan, 2001). Often, the triggers associated with SCD events are physical exertion (as seen in LQT1), auditory stimuli (as seen in LQT2), and sleep/rest (in LQT3). While the likelihood of a cardiac event is highest in patients with LQT1, attacks that can result in sudden cardiac death are often in patients with LQT3 (Khan, 2001).

**Treatments for long QT syndrome.**

Treatment for long QT syndrome is critical for both symptomatic and asymptomatic patients. When left untreated, mortality rates increase drastically. Statistics show that more than 20% of those patients who are symptomatic and do not receive treatment die within 1 year of first episode of syncope, and nearly 50% die within 10 years of diagnosis. When properly treated, the 5-year mortality rate decreases to 3% to 5% (Chiang, 2004). Clearly, proper treatment is crucial for long-term survival. Management
of LQTS primarily consists of three different treatments used independently or in conjunction with each other. These treatments consist of beta-blockers and lifestyle changes, cardiac pacing or implanted cardioverter defibrillators (ICDs) and left cardiac sympathetic denervation (LCSD) (Chiang, 2004; Collins & Van Hare, 2006; Goldenberg & Moss, 2007; Kapetanopoulos et al., 2006; Kass & Moss, 2003; Khan, 2002).

**Medications.**

Beta-blocker medications have become a primary therapy for patients with LQTS. Beta-blocker therapy significantly decreases the likelihood and frequency of cardiac events. However, this therapy does not completely eliminate the dangers of sudden cardiac death. While beta-blockers remain most widely used treatment, different types of LQTS respond differently to this type of medication. Beta-blockers are very successful in treating patients with LQT1; however, they offer less protection to persons with LQT3 (Chiang, 2004; Collins & Van Hare, 2006; Goldenberg & Moss, 2007; Kass & Moss, 2003; Khan, 2002). Beta-blockers, such as propanolol, are used to protect persons with LQTS from experiencing torsades de pointes, thus protecting them from cardiac events and SCD. However, the use of beta-blocker medications may be ineffective, and patients may still be symptomatic while taking this medication. Magnesium and potassium supplements are often used in this population. Both of these elements can be used to treat acute episodes of torsades (Khan, 2002).

**Implanted cardioverter defibrillators and pacemakers.**

Implanted cardioverter defibrillators (ICDs are the most effective treatment for high-risk LQTS patients (Chiang, 2004; Collins & Van Hare, 2006; Daubert et al., 2007; Goldenberg & Moss, 2007; Kapetanopoulos et al., 2006; Kass & Moss, 2003; Khan,
ICDs are used in conjunction with beta-blockers, not as an alternative to beta-blocker medications in symptomatic patients, but to protect them from arrhythmias that are not prevented by the medications. The use of ICDs has reduced the mortality rate from 14% over 8 years to 1.3% over 3 years (Chiang, 2004).

The success of the implantable cardioverter defibrillator for patients with LQTS has increased its use as a treatment option. ICD implantations are being conducted more often now as both a primary (no cardiac event/high risk-patient) and secondary (after aborted SCD) treatment for LQTS (Collins & Van Hare, 2006). However, the implantation of an ICD does not come without risks, both physical and emotional. There are concerns with device-related issues such as lead fractures, inappropriate shocks, and device malfunctions. Further, there are many psychological issues that can result from the implantation of an ICD. These issues include symptoms of anxiety after appropriate or inappropriate shocking and body image issues. In addition, the implantation of such a device can cause emotional arousal, which can trigger arrhythmias and result in repeated ICD shocks, called electrical storm (Chiang, 2004). The success of ICDs in preventing sudden cardiac death in patients with LQTS makes them an important treatment for LQTS.

Pacemakers are also used to treat LQTS patients. The pacing rates can be finely adjusted in these devices to prevent lower heart rates that results in pauses that can trigger torsades de pointes (Khan, 2002). Primarily, cardiac pacing is used in conjunction with beta-blockers in patients with bradycardia, or slow heart rates (Chiang, 2004). LQT3 patients benefit from such treatment because they are at risk of having bradycardia during sleep, while they are in their most vulnerable state.
**Surgical left cervicothoracic sympathetic denervation.**

Surgical left cervicothoracic sympathetic denervation (SLCD) is a surgical procedure used for patients who experience repeated syncope and ICD firings despite their regimen of beta-blocker medications (Chiang, 2004; Goldenberg & Moss, 2008; Khan, 2002).

**Lifestyle modification.**

Lifestyle modification is an important aspect of treating patients with LQTS. The delicate nature of their hearts’ electrophysiology requires that these patients conform to restrictions in their activities and lifestyles. For instance, strenuous exercises in general are to be avoided in patients with LQTS (Maron et al., 2004). Kapetanopoulos et al. (2005) provide information on the acceptable levels of different sports and recreational activities for patients with LQTS. However, patients with LQT1, in particular, should avoid strenuous, competitive activity, especially swimming (Chiang, 2004; Collins & Van Hare, 2006; Kapetanopoulos et al., 2005; Napolitano, Bloise, & Priori, 2006). On the other hand, patients with LQT2 are at a lower risk of syncope or SCD during exercise and exertion. Lifestyle modifications in this expression of LQTS require that marked emotional stress, startling, and auditory triggers such as loud noises be avoided. These patients should switch to phones that vibrate, avoid the use of alarm clocks, and be very cautious of their environment. Such concerns would include school bells, church bells, loud movies, fire alarms, sirens, and other loud and sudden noises (Chiang, 2004; Kapetanopoulos et al., 2005). Therefore, the type of LQTS dictates the treatments required to keep individual patients from experiencing the severe symptoms of LQTS.
Treatment adherence.

Treatment adherence and treatment compliance have been an area of concern within the medical community for thousands of years. Medication adherence and issues with compliance have been dated back to the time of Hippocrates. At that time, the effectiveness of a potion was evaluated by making a record of whether or not patients were taking the prepared treatment. Hippocrates, in his own medicinal research, was aware that some of his patients were not adhering to his prescribed medications. Things have not changed much, and the concern surrounding treatment adherence continues to be an obstacle in proper illness management. In chronic illnesses such as long QT syndrome, treatment adherence is critical for the prevention of potential fatal symptom expression (Osterberg & Blaschke, 2005; Turchin, Kolatkar, Pendergrass, & Kohane, 2007).

Definitions of compliance and adherence.

Treatment adherence or compliance has been generally defined as the degree to which a patient follows a prescribed treatment protocol as outlined by their health care provider. Adherence is the preferable term used by physicians, as it implies a therapeutic alliance and an agreed upon treatment plan or course. The term compliance is believed by some to imply that the patient is a passive member in his or her treatment team and simply follows the physician’s orders without any of their own input. Furthermore, compliance is sometimes framed as representative of an authoritarian patient-physician relationship, in which the patient is expected to obey orders from their health care provider, implying a subservient role. The term adherence is preferred to compliance because of the negative connotations associated with the latter (Dezii, 2000;
Osterberg & Blaschke, 2005; Sultan, Bungener, & Andronikof, 2002). For the remainder of this paper, the terms compliance and adherence will be used interchangeably without an intended negative connotation.

Sultan, Bungener, and Andronikof (2002) remark that nonadherence is an idea that can only be defined using its opposite, adherence. This is part of the reason that clinicians have such difficulty defining these terms. In order to have a strict definition of nonadherence, one would need to have a strict definition of adherence. A problem in the literature is that there is no operational definition of these terms. Sultan and colleagues define adherence as the tendency for a patient to comply or follow the advice of their health care provider in regards to their treatment and regimen. Cramer et al. (2008) define compliance as the degree to which a patient conforms to recommendations of their treatment provider with specific respect to dosing, time, and frequency. Therefore, the research that makes an effort to understand and treat noncompliance and nonadherence lacks consistency and has failed to make a great contribution to the increasing problem of treatment nonadherence (Cramer et al., 2008). In fact, there has been a large amount of research on the subject of nonadherence/noncompliance; however, little change has resulted. Furthermore studies suggest that, despite research of etiology and intervention for treatment noncompliance, health care interventions have limited efficacy for increasing adherence (Haynes, Yao, Degani et al., 2005; McDonald, Garg, & Haynes, 2002).

**Consequences of noncompliance/nonadherence.**

The consequences of noncompliance are considerable. Noncompliance impacts the economy, physicians, hospital administrations, and individual patients. It is estimated
that treatment noncompliance costs the United States government approximately $100 billion each year, of which $30 billion are in direct costs and $25 billion are due to hospital admissions (Elliott, Shingole, Peele, Bhosie, & Hughes, D. 2008). Furthermore, 33% to 69% of all medication-related hospital admissions are due to complications of nonadherence (Dezii, 2000; Osterberg & Blaschke, 2005). Noncompliance with treatment also results in increased illness and mortality. The World Health Organization (WHO) indicates that despite the well-documented efficacy of treatment adherence in reducing symptoms and increasing health, long-term adherence rates are only 50%, regardless of illness or treatment regimen (Lehane & McCarthy, 2007; Young & Oppenheimer, 2006). Furthermore, approximately 10% of hospital admissions and 23% of nursing home admissions each year are due to medication noncompliance (Young & Oppenheimer, 2006). Nonadherent behavior is seen in all illnesses, both chronic and acute. Two examples of the impact noncompliant behavior has on patients can be understood by looking at research related to diabetes and asthma.

*Noncompliance in diabetes.*

Diabetes is a leading cause of death in the United States, contributing to more than 190,000 deaths each year. The number of persons in whom diabetes is diagnosed is dramatically increasing each year. Diabetes is a chronic illness that requires preventative and management efforts to ensure the best quality of care for patients. Persons with diabetes typically have a unique and personalized treatment regimen provided to them by their physician. Proper diabetes care requires strict adherence to these individualized plans. With proper management, persons with diabetes can live relatively normal lives (Helme & Grant Harrington, 2004; Hill-Briggs, 2003).
Proper diabetes management includes adhering to food limitations, physical activity, medications (including insulin), monitoring and adjusting blood glucose levels, and seeking appropriate medical attention when necessary. Treatment adherence in diabetes is critical. Failure to comply with personalized regimens and maintenance programs can result in severe side effects, such as loss of vision, limb amputation, coma, or even death. There are currently more than 20 million people in the United States with diabetes, and it is estimated that between 50% and 80% of diabetics are noncompliant with treatment (Chatterjee, 2006; Hill-Briggs, 2003).

Children and adolescents with diabetes face their own unique resistance to the diagnosis and its adherence requirements (Dantzer, Swendsen, Maurice-Tison, & Salamon, 2003; Weinzimer, Doyle, & Tamborlane, 2003). Mulvaney et al. (2008) examined the barriers to treatment adherence that are specific to youth. It was indicated that the predominate barriers to proper self-management and compliance in adolescents with type 2 diabetes included psychosocial development and comfort, others with diabetes (family/friends), environmental factors, and their coping skills and problem-solving abilities. Interference with proper compliance was in part the result of adolescents trying hard to be “normal” and fear of being embarrassed of their dietary restrictions and medication requirements. Further, the role of the adolescent’s family on compliant behavior is greatly impactful. Many of these youngsters (typically with type 2) have family members who share this diagnosis. Positive role modeling may encourage adherent behavior while negative role modeling may encourage nonadherent behaviors (Mulvaney et al., 2008).
Environmental factors also influence youth with diabetes, with respect to proper self-management of their blood glucose levels. Many adolescents encounter barriers to proper treatment. Examples of these include finding appropriate foods at school and while spending time with peers outside of school, feeling as the need to isolate themselves to test and manage their blood glucose levels, and needing to leave class early for medication or treatment. The ability to effectively cope and overcome these barriers is necessary to increase compliant behavior. Educating their peers and including them in their management requirements, learning effective communication skills with parents, peers, and medical staff to voice concerns, and planning behaviors are methods proposed to increase compliance in this population (Mulvaney et al., 2008).

*Noncompliance in asthma.*

Asthma affects nearly 20 million Americans and results in nearly 4,000 deaths annually. While there is no cure for asthma, management through several therapies is used to control symptoms and attacks. Suboptimal control of asthma symptoms is a common result of poor treatment adherence. While there are no statistics on the rate of noncompliance in this large population, research has determined that compliant patients are significantly less likely to have acute asthmatic attacks and exacerbations of symptoms (Stern et al., 2006; Walewski, Cicutto, D’Urzo, Heslegrave, & Chapman, 2004).

Asthmatic patients may struggle with compliance for several reasons. It is suggested that these reasons include dosage frequency, the slow onset of action, and the questionability of future adverse affects. The pediatric population also experiences these difficulties. Colland, van Essen-Zandviet, Lans, Denteneer, Westers, and Brackel (2004),
determined that pediatric asthmatics may have increased problems with compliance due to the illness beliefs that they hold. Illness beliefs, such as the belief that their symptoms and difficulties will disappear, influence compliance through thoughts of denial and avoidance. Further, children with little understanding of the cause of symptom exacerbation also had lower adherence rates. This data speaks to the importance of providing accurate information to children about their diagnosis and management (Colland et al., 2004).

**Measures of adherence.**

As previously discussed, a significant obstacle in understanding adherence challenges and developing interventions for treatment nonadherence is the lack of an operational definition. Health care providers cannot agree on what defines a missed dose or what the proper action should be if a missed dose or nonadherence is identified. For example, in a qualitative study by Sankar, Nevedal, Neufeld, and Luborsky (2007), 35% of clinicians included a delay in taking medications as noncompliance, 41% of clinicians believed late administration of medication is part of noncompliance, and 12% stated that late ingestion of medications is only a noncompliant behavior if over 24 hours late, while 18% believed it is the next dosing time that defines late ingestion. Clearly, there is little consensus. For example, taking medications 4 hours late would be considered noncompliance by only some physicians. However, even with no consensual standard of measure for what noncompliance means, it is still important to identify methods by which researchers determine if noncompliance is taking place (Sankar et al., 2007).

In regards to measures of adherence, there are two methods or modes of adherence; direct methods and indirect methods. There are also direct and indirect
methods of measurement for quantifying medication compliance. Direct methods include measuring concentrations or metabolization of medications in blood or urine. These approaches tend to be expensive and laborious. In these examinations, subtherapeutic levels of medications imply poor adherence management (Osterberg & Blaschke, 2005). An LQTS patient being monitored for medication compliance directly would have routine blood tests to determine if therapeutic levels are present.

Indirect methods of measuring adherence include asking the patient about his or her medication taking habits. Such methods include asking questions about the ease of the program, pill counting, rates of prescription refills, electronic medication dispensers, diary/journal records, etc. However, there is limited reliability in each of these methods of measurement, resulting in health care providers overestimating compliance rates. For example, pill counting is a very common measurement tool; however, patients can switch medications or discard them entirely. In terms of digital or electronic monitoring systems, these systems are excellent at monitoring behaviors surrounding medication taking (e.g., time compliance, refilling prescriptions); however, they cannot document whether the medication in question was ingested after being dispersed. In addition, such devices are not covered by insurance, quite costly, and not routinely used (Osterberg & Blaschke, 2005).

Indirect measurement of LQTS compliance could include asking the patient about his or medication compliance as well as about their level of activity, avoidance of activities (swimming), sleep patterns, etc. Medical professionals can use prescription refill rates to determine if LQTS patients are taking proper doses of medications.
Health belief model.

The health belief model (HBM) is an organizing framework used to understand, explain, and predict the way people accept changes in their health and their acceptance of recommendations made by medical professionals (Janz & Becker, 1984). As a psychosocial model developed to understand compliance and adherence trends in patients, the HBM examines the value patients place on a goal and the belief or the likelihood that they can achieve it. It is for this reason that the HBM is also referred to as a value expectancy model (Harrison, Mullen, & Green, 1992). The health belief model has been found to be a reliable method for understanding compliance and adherence complications in various chronic illnesses, including renal diseases, hypertension, asthma, obesity, osteoporosis, middle-ear infections, and other chronic conditions (Becker & Janz, 1984; Bush, 1990; Roden, 2004).

The health belief model was developed in the early 1950s by a team of social psychologists lead by Dr. Rosenstock at the U.S Department of Health and Human Services as a method to understand treatment nonadherence and the nonacceptance of preventive medicine. The HBM is based on principles, including motivation, attitude, and perceptions of health, of both cognitive behavioral therapy and learning theories. The HBM is often used to understand preventative health behavior (or the lack of such behaviors), which directly connected to treatment adherence and compliance concerns as well (Bloom-Cerkony & Hart, 1980; Janz & Becker, 1984; King, 1980).

The health belief model is an empirically supported framework of understanding treatment adherence (Becker & Janz, 1984; Bloom-Cerkony & Hart, 1980; Janz & Becker, 1984; King, 1980). The HBM is based on the assumption that health is a
commodity that is highly valued and a highly ranked goal for individuals. However, as a psychosocial approach, the HBM does not specifically dictate any intervention protocols or strategies. However, the HBM is practical in the clinical setting because of its patient-centered approach to medicine. Using the HBM, medical professionals can use information regarding patients’ perceptions, attitudes, and beliefs about treatment compliance and be more flexible regarding individual needs when providing treatment options. For example, a physician may allow a child to play soccer if he/she plays as a goalie in order to reduce periods of extended exertion.

Patients’ acceptance of medical advice or information is often influenced by their subjective beliefs and level of understandings about medical protocols. Without this understanding of individual perceptions, attitudes, and beliefs, medical professionals would not know what concessions or negotiations in treatment can be made to increase adherence and improve the health of their patients. The HBM can assist both physicians and patients in better understanding their limitations and areas of growth in medication compliance (Becker & Janz, 1984; Bloom-Cerkony & Hart, 1980; Gillibrand & Stevenson, 2006; Janz & Becker, 1984; King, 1980).

Health belief model: Dimensions.

As previously stated, the health belief model is an empirically supported framework for understanding medication and treatment compliance in chronically ill patients. The health belief model consists of individual perception variables, as well as modifying variables that influence an individual’s choice or tendency to act. A patient’s perception of the situation is impacted by modifying variables such as demographics, personality
styles, social class, education, and prior disease knowledge. These factors influence the way in which the HBM serves to work (Ali, 2002).

The HBM is founded on two primary variables: first, the degree to which a person places value on a particular goal, and second, an estimate of how likely a given action will enable him or her to attain that goal. These variables are further explored from a medical standpoint and are redefined with an emphasis on health-related behaviors. These more specific definitions are, the desire to avoid illness, or the value placed on health, and the belief that a health behavior will prevent illness (Becker & Janz, 1984; Bond, Aiken, & Somerville, 1992; Janz & Becker, 1984). A person with LQTS would define this as the desire to avoid syncope or sudden cardiac death and the degree to which they believe medications, activity restrictions, and various surgical procedures will help to prevent these potentially fatal events. The HBM is comprised of four dimensions. They are perceived susceptibility, perceived severity, perceived benefits, and perceived barriers.

*Perceived susceptibility.*

Perceived susceptibility is the degree to which individuals feel vulnerable to a condition or their perception of perceived risk of contracting or having an illness. Also in this dimension is the perceived risk of resusceptibility to a condition or illness state. Resusceptibility includes illness reoccurrences or, in the case of LQTS, symptom flairs (e.g. syncope, fatigue) or the experience of sudden cardiac death (Janz & Becker, 1984).

*Perceived severity.*

Perceived severity is an individual’s feeling or belief on the seriousness or severity of the medical complication. This includes personal evaluations of medical consequences
(e.g., pain, death) and social consequences (Janz & Becker, 1984). The level of perceived severity in LQTS patients would be the degree to which they believe LQTS to be fatal.

**Perceived benefits.**

Perceived benefits include an individual’s feeling or belief that their efforts in treatment are worthwhile and efficacious. This dimension is formulated on the idea that patients would not undergo and comply with treatment regimens if they did not believe their efforts to be both feasible and effective (Janz & Becker, 1984). In LQTS patients, the perceived benefit of treatment would be the degree to which they believe their medications, activity restrictions, and possible surgical procedures are aiding in reducing symptoms and ultimately saving their lives.

**Perceived barriers.**

The barriers that are perceived by patients are the potential negatives of engaging in health-promoting behaviors. These perceived barriers negatively affect health by impeding compliance with recommended treatment. Examples of perceived barriers include but are not limited to cost, side effects, pain, restrictions, and consumption of time. A noncompliant LQTS patient may identify the perceived barriers of treatment as overwhelming. They may believe, for example, that having to quit the soccer team and have surgery to implant an ICD is too painful, too invasive, and too restrictive of their quality of life. Another example would be a patient who feels that the fatigue experienced from beta-blockers is not worth remaining medication compliant. Each of these health belief variables or dimensions influences both the decision to act and the
Several studies examining the health belief model in conjunction with illness, including diabetes and coronary heart disease, found that the four dimensions outlined above were not equal in their influence in making medical decisions. In each case, perceived susceptibility accounted for the largest and most significant predictor of compliance and positive health behaviors. However, during acute illness or acute onsets of symptoms, perceived severity became a better predictor of adherence. Studies examining preventative health behaviors found barriers to be a more influential dimension in health-related decision making (Becker & Janz, 1984, Bloom-Cerkony & Hart, 1980). LQTS often requires preventive measures to avoid syncope and sudden cardiac death. In LQTS, perceived barriers may be a more influential dimension in determining compliance because of the preventive nature of many of the restrictions and treatments that are made. Persons with LQTS may minimize the severity and susceptibility of illness and increase the value of limitations.

Rosenstock devised these dimensions to be complimentary and balanced. Higher perceived severity and susceptibility cause patients to be more alert and possibly more proactive in treatment, while perceived barriers and perceived benefits provide patients with a path or avenue from which they may choose to act or not act. Rosenstock notes, however, without a trigger, or cue to action, the decision-making processes would fail to launch (Janz & Becker, 1984).
Illness attributions and the health belief model.

King (1983) brought to light the importance of illness attributions and their impact on illness perception in the health belief model. Illness attribution is defined as the explanation of the origin of illness, or the belief about why a person has a particular illness and/or what they have or what is “wrong” with them. These illness attributions are very subjective and highly individual to the patient and their perception and understanding of the illness (King, 1983).

Literature on illness attributions divides illness attributions into two categories. The first category is the locus of control category. This grouping of attributions characterizes persons who hold the belief that the outcome of their (medical) behaviors is under their control. This group is more internally oriented and feels more personal control over their general health and well-being. A patient with LQTS with an internal locus of control may view their medication consumption or activity restriction, for example, as proactive behaviors to best ensure that they avoid cardiac events. The second category of research has focused on specific health information and explanations of illness based on compliant and preventative behaviors, as well as on coping strategies. In other words, this research has examined how individuals attribute the cause of events or behaviors. When attributing the cause of illness to one’s self, coping appears to be stronger due to the adaptive functioning of gaining control over what would otherwise be inexplicable. However, research also shows that attributing the cause of illness to oneself can have maladaptive consequences, such as self-blame, guilt, and depression. This type of expression may be seen in those few patients who develop acquired LQTS. The way in
which a patient attributes the cause of illness (internal vs. external) can influence their decision to seek medical care and affect accurate reporting of symptoms (King, 1982).

Causal attributions are another factor to be considered when looking at the effects attributions have on health behaviors. Traditionally, causal attributions have been differentiated as being internal and external. However, Kelley (1975) suggests that individuals use three different types of information to make causal attributions. These sources of information are consensus, consistency, and distinctiveness. Consensus information relates to the degree to which other people react in a common way to an illness. Consistency of information is the degree to which an individual’s behavior matches his or her behavior in similar situations. Finally, distinctiveness information is the degree to which a person responds in a similar way to other varying situations (Kelley, 1973). Persons with LQTS may have causal attributions that increase or decrease their likelihood of being treatment adherent. Persons with LQTS and their caretakers who are proactive and optimistic in their daily lives may be able to generalize this style of thought to the preventive care required for proper treatment of LQTS. This consistency, rather than distinctiveness in attribution, would tend to make this patient treatment adherent. King (1983) hypothesized that illness attributions, causal attributions, and health beliefs all impact and influence health-related behaviors. Clinicians can use patients’ health beliefs and causal attributions to predict treatment-related behaviors (King, 1983).

The way in which a patient attributes their illness can also be linked to and predict adherence to treatment. Illness attributes influence behavior in two ways. The attributions affect health beliefs and consequently health-related behaviors.
attributions can directly impact behavior. However, in both pathways, an individual’s personal health beliefs, in addition to his or her illness attributions, result in decision making and ultimately health-related behaviors.

**Health belief model and treatment adherence.**

The health belief model is influential in understanding treatment adherence behavior. Many chronic illnesses, including LQTS, require patient adherence to maintain effective control and prevent potentially fatal events. Medical professionals are often surprised by the high levels of noncompliance found in patients (Becker & Janz, 1984). However, when patient care and compliance are viewed from the perceptive of the patient, the reasons for treatment nonadherence become clearer. The HBM is one such way that we can better understand chronic illness compliance from the perspective of the patients. On the whole, compliant patients perceive their illness as more serious than noncompliers (Becker & Janz, 1984; Bloom-Cerkoney & Hart, 1980; Bond et al., 1992). In addition, treatment compliers tend to experience more symptoms than do noncompliers. Thus, persons with LQTS who experience frequent syncope, fatigue, or appropriate/necessary firing of their AED/ICD will most likely be more compliant than the LQTS patient who does not experience any symptoms. Conversely, the complexity, duration, and rate of change that treatment requires, and the degree to which treatment impedes daily functioning, reduce compliance levels (Bloom-Cerkoney & Hart, 1980).

In addition, severity and susceptibility increase compliance, while perceived barriers decrease compliance (Bloom-Cerkoney & Hart, 1980; Bond et al., 1992). Cues to action are also an important ingredient of treatment adherence, as found by Bloom-Cerkoney and Hart (1980), as they are responsible for triggering compliant behavior.
Cues to action can be external events such as alarms, reminders from family, or predetermined times (e.g., medications are taken at 10:00 AM and 10:00 PM). Likewise, cues to action can also be internal and would include triggers such as not feeling well. These cues to action are considered in this model and are important to compliance issues because they often trigger compliant behavior (Patino, Sanchez, Eidson, & Delamter, 2005).

**Theory of reasoned action.**

The theory of reasoned action (TRA) was developed by Fishbein and Ajzen. The theory of reasoned action was developed as a method to explain the relationship between attitudes and behavior (Anderson & Lavallee, 2009; Clarke & Becker, 1998; Fishbein & Ajzen, 1975; Romano & Netland, 2008). The TRA conceptualizes an action, such as taking prescribed medications, as the person’s willingness to perform a behavioral intention. The intention or behavioral intention, as used here, is influenced by an individual’s attitude toward performing the behavior, as well as his or her belief of the subjective norms of that desired behavior. A person’s attitude is determined by the perceived consequences of performing a behavior. In terms of LQTS and treatment compliance, this can be translated into a person’s willingness to take prescribed medications or adhere to physical restrictions as prescribed by the physician, in conjunction with his or her personal attitude and belief about the degree to which compliance behavior will be of benefit and how family/friends/the medical team will perceive their compliant or noncompliant behavior. The TRA states that an individual’s attitudes and perception of norms impact behavioral intentions, thus impacting behavior
(Anderson & Lavallee, 2009; Fishbein & Ajzen, 1975; Romano & Netland, 2008; Sultan, Bungerner, & Andronikof, 2002).

**Attitudes and subjective norms.**

The theory of reasoned action theorizes that attitudes and norms do not influence or determine behaviors directly. Instead, attitudes and norms influence a person’s behavioral intention. Fishbein and Ajzen (1975) define behavioral intention as the probability that a person will act or behave in some way. Intention is consists of four elements: the behavior, the target or object of the behavior, the situation in which it was performed, and the time when the behavior will be executed. These four elements in a LQTS patient would include a patient’s intention to take the appropriate medications at specific times, as indicated by the medical team. According to the TRA, the behavioral intention to take medications or adhere to other medical advice is influenced by the patient’s attitude towards the behavior, as well as the subjective norm he or she understands surrounding the behavior (Fishbein & Ajzen, 1975). If, for example, an LQTS patient was medication and treatment compliant soon after diagnosis but experienced several events of syncope, he or she may have the attitude that being properly medicated will not prevent these unexpected events. Thus, according to this patient, the consequence of not performing the behavior (being compliant to treatment) is minimal. Furthermore, they may have grown up in a family/culture that did not rely on conventional medication and preferred to use diet and exercise as a method of prevention and proper self-care. This patient’s attitude toward being treatment compliant would be negative, due to early experiences and subjective norms that influence nonconventional
medical decisions. This patient, regardless of initial intention, may not be treatment compliant.

Behavioral intentions.

Behavioral intention is a function of both the attitudinal beliefs held by an individual and his or her understanding of the subjective norm. The predictability of behavioral intention varies in relation to the complexity of the behavior. The intention to engage in a single behavior is more predictable than the intention to engage in a complex string of behaviors. For example, the intention to be treatment compliant all day for a single day is more predictive of such behavior than the intention to be completely adherent all year. In summary, the likelihood that a person engages in a behavior is determined by his or her intention at that point in time (Fishbein & Ajzen, 1975).

Stabilizing a person’s intention then becomes the next area of interest in this model. Intentions to act are not stable, and can vary by hour, day, week, or year. Treatment adherence is critical to symptom management and survival and requires that a high degree of behavioral intention to adhere to treatment remains stable over time. Fishbein and Ajzen state that the longer the interval between the behavioral intention and the execution of the behavior, the lower likelihood that the behavior will be executed. The stability of intention can be influenced by changing attitudes and beliefs as well as a change in perception of the subjective norms. This speaks favorably for the ability to correct noncompliance in patients. The medical team can offer information and support in an effort to change the patient’s attitude toward medications and convey their expectations of the patient and the expectations of the medical field to alter the patient’s subjective norm of treatment nonadherence (Fishbein & Ajzen, 1975). A patient with
long QT1, for example, who is engaging in nonadherent behaviors, such as medication noncompliance and swimming on a regular basis, could have his or her attitude toward these behaviors altered by talking to the medical staff, fellow patients, and mental health consultants to better understand the degree of risk they are taking. Medical staff can impart their expectations of compliance, and other LQTS1 patients can state their beliefs about and standards of treatment adherence in an effort to shift the subjective norm that the nonadherent patient holds.

Another area of intention stabilization that may interfere with younger patients with LQTS is the degree to which intention to act is dependent on someone else. For younger persons with LQTS, they may rely on their parent or guardian to provide the proper care. Children may not intend to be noncompliant; however, their behavior is noncompliant. If a person’s behavior is based on expectation that another person will perform in some way their behavior may not match their intention (Fishbein & Ajzen, 1975).

According the theory of reasoned action, the intentional behavior is an antecedent to the behavior itself. However, this seemingly simply concept is complicated by intervening issues, such as varying attitudes and beliefs and evolving subjective norms (Fishbein & Ajzen, 1975). Such intervening variables influence both the intentional behavior and the resulting behavior. For example, a family with LQTS may have the intention to be compliant with their medications; however, the breadwinner may lose his or her job and health insurance. This previously adherent family may decide that they can only afford half of the medications and choose to medicate their child(ren) with LQTS, rather than the parent with LQTS, until they can get new insurance. However,
Fishbein and Ajzen and their TRA model determined that behavioral intentions with minimal intervening variables are a good predictor of behavior (Fishbein & Ajzen, 1975).

**Changing intentions and behaviors.**

The theory of reasoned action can be used to conceptualize a method of change. As previously explained, this model uses an individual’s attitudes toward or beliefs about a behavior and his/her understanding of the subjective norms associated with that behavior in deciding the degree to which he or she intends to act. Therefore, the ability to change or influence the level of intention to behave in a certain way, such as being treatment adherent, is dependent on the attitudinal and normative understandings of the action. Initiating change would then be dependent on changing attitudinal and normative understanding of the behavior. Cognitive behavioral therapy (CBT) is an excellent mechanism by which to challenge and reconstruct maladaptive assumptions and beliefs about treatment adherence behaviors. Using thought records and cognitive restructuring, a therapist would work with a nonadherent patient to alter his or her perception of treatment adherence and have him or her reframe normative misconceptions (Beck, 1995; Fishbein & Ajzen, 1975).

**Medication adherence model.**

The medication adherence model developed by Johnson (2002), was developed from a qualitative study examining 21 older adults with hypertension and their adherent and nonadherent behaviors as related to their intention and unplanned interferences in medication management of their condition (Johnson, 2002; Lehane & McCarthy, 2007). The medication adherence model, also known as MAM, is comprised of three main concepts: purposeful action, patterned behavior, and feedback. Purposeful action is the
level of intention a person has to be compliant to treatment based on their understanding of their need, outcome, and safety of adherence. Patterned behavior refers to a patient’s routine and the initiation of developing a personal treatment routine or pattern of taking medications. Lastly, feedback on compliance includes the information or facts they receive about their treatment regimen (Johnson, 2002; Lehane & McCarthy, 2007).

The MAM is driven by the idea that persons are noncompliant for two dominant reasons: the intentional decision to be noncompliant and unintentional interferences with their compliance regimen. Lehane and McCarthy (2007) both conducted and examined additional studies to determine the utility of this model. Results of their investigations found that the vast majority of noncompliant patients report unintentional nonadherence (31%) versus intentional noncompliant behavior (9%). These results can be useful in conceptualizing the obstacles to treatment compliance, specifically medication compliance. These findings suggest that behavioral interventions may be useful in preventing unintentional nonadherent behavior. For example, a family member with LQTS may choose to have his/her medications in a visible and accessible place. Therapists could implement behavioral interventions such as stimulus control. Stimulus control refers to a change in behavior due to the presentation of a discriminative stimulus. The presentation of discriminative stimuli increases the probability that an organism will respond (Franks, 1984; Pierce & Cheney, 2004). Placing medications on the bathroom sink could trigger a person to take medications each morning and evening as part of their established morning and evening routine, such as brushing their teeth.
Summary.

In summary, medication adherence is critical to the successful management of long QT syndrome. Without proper management of LQTS, patients may experience symptoms such as arrhythmias, syncope, and/or sudden cardiac death. Treatment adherent behavior increases the likelihood that a person with LQTS can live a relatively normal and long life. As a growing number of individuals are being screened for LQTS and other sudden cardiac arrest diseases, increases, LQTS is being diagnosed more frequently. This increase in population requires treatment teams to find ways to identify and reduce barriers to treatment adherence.

There is currently little research addressing the psychological effects of having a LQTS and the impact of these psychological effects on treatment adherence. LQTS, like other chronic illnesses, directly affects the daily functioning of those who have it. It is therefore in the best interest of physicians and treatment teams as a whole to understand and consider the psychological ramifications of such an illness and the influence this has on management. The purpose of this study was to increase awareness of the psychological effects of LQTS, as well as to examine personal struggles with treatment adherence, in an effort to develop a model of understanding treatment adherence within the context of LQTS.

Research question.

The research question utilized to guide this study was: What are the developmental, social, emotional, and behavioral factors experienced by persons with long QT syndrome that affect their treatment adherence?
Chapter 3

Methods

A qualitative, grounded theory design was used to explore risk factors and behaviors related to treatment noncompliance in persons with long QT syndrome. Participants were persons who contributed to an online message board that is designed to offer support to families and persons with LQTS. The following chapter will examine methodology, participant requirements, procedures used in this study, and an objective determination of the study’s utility in the growing field of clinical health psychology.

The design chosen for this study was a qualitative, grounded theory design. Qualitative research is often used for areas of study that are in the early stage of exploration and would benefit from a broad exploratory study. Psychosocial research regarding persons with long QT are sparse at best; thus, qualitative methodology is a preferred method to explore psychological effects surrounding long QT and treatment adherence. Qualitative methodology can generate further psychosocial research on LQTS and benefit this growing community.

Grounded theory design methods were chosen as a specific methodology. This approach builds theory from data. Grounded theory allows researchers to extract theory from collected data rather than searching for their hypothesis to emerge from the data. The flexibility of grounded theory allows researchers to fully examine individual behavior and social phenomena with a nonbiased approach (Corbin & Strauss, 2008; Dolan Mullen, 2006).

In addition, many techniques of qualitative research were used in the analysis of the data. These techniques included the use of questioning, the flip flop technique, waving
the red flag, looking at language, looking at emotions that are expressed, “so what?”
“what if,” and looking for the negative case. Each of these techniques allows for more a
more in-depth and analytical view into the data. These tools are fundamental to such
analysis and add to the degree to which researchers can extrapolate from the data (Corbin
& Strauss, 2008).

The advent of the Internet has increased the mechanisms by which members of the
sciences can collect and analyze data. This study utilized computer-mediated
communication (CMC) to conduct a qualitative study, instead of the more common face­
to-face methods (FTF). Computer-mediated communication allowed this study to
transcend geographical boundaries and obtain a sample that is international. Utilizing
CMC allowed the researchers to reach a specific population and retrieve information
from more than 780 possible subjects. If FTF communications methods were used to
communicate with this large number of subjects, the time and costs associated would be
astronomical (Mann & Stewart, 2000).

Subjects in this computer-mediated communication were not aware of their
participation in the study and therefore there was no concern that they were censoring
their information. According to Eysenbach and Wyatt (2002), the material on the
Internet can serve as an expansive and valuable resource to investigators interested in
qualitative research. The internet provides researchers with unique accessibility to
information with complete anonymity. Eysenbach and Wyatt (2002) expressed the
unique nature in which the internet can serve as a medium to gain qualitative information
about groups. They stated, “in qualitative research we are not interested in an average
view of a patient population, but want to gain an in-depth understanding of the
experience of particular individuals or groups” (Eyssenbach & Wyatt, 2002). It is important to consider that there are limitations to Internet-based research. One such limitation is that the limited number of persons who have access to the Internet (because of financial ability, time, and/or literacy) are the only individuals represented in the research, limiting generalizability.

There was no potential risk to subjects because confidentiality was maintained. There was no direct benefit to the subjects in this study. The potential benefit of this study is to the long QT population in general, as well as to persons with other chronic illnesses.

Subjects.

Subjects in this study were persons chosen from a long QT syndrome online message board group who contributed to the long QT syndrome message boards between February 1, 2008, and February 28, 2008, and between September 1, 2008, and Sept 30, 2008. All entries in this period were analyzed as part of a larger qualitative study on psychosocial factors relating to LQTS. There are approximately 780 international members on the long QT message board. All persons in the online user group are impacted by long QT (directly or indirectly) and were included in this larger study on long QT syndrome. For this individual study, threads were included based on their relevance to treatment adherence/compliance and were excluded if they did not mention treatment adherence concerns.

Screening procedures for inclusion and exclusion criteria.

Screening procedures used for inclusion and exclusion consisted of the topic of subject participation. This study included only those subjects who discussed issues of
medication noncompliance and excluded all threads that did not support this topic of exploration, as determined by three to four individual coders and their consensus on this classification.

**Recruitment.**

This study did not require direct recruitment. The message board entries were archived by the user group. Subjects were unaware that this study was being performed. It was determined that this was the best course of action because notifying this population of this study may have influenced participation in an unfavorable way and negatively affected their support network by contaminating the group. The group moderators were informed and gave permission for the use of data for this research.

**Procedure.**

The current study was part of a larger qualitative study of persons with LQTS. Messages were downloaded and deidentified (if necessary) prior to this research and analysis of the data. In an effort to maintain confidentiality, subjects were not identified to the researcher or coders by their names. Subjects are identified on the listserv by their e-mail address rather than by their legal name. Because e-mail addresses could compromise confidentiality, the researcher identified each subject by a unique code. No identifying information was used in the analysis of messages or discussion of the analysis. All data was deleted at the conclusion of the study.

Messages between February 1, 2008, and February 28, 2008, and between September 1, 2008, and September 30, 2008, were chosen to avoid major holidays, but with seasonal differences in mind.
There were seven individuals involved in the coding of the data. Five of these individuals are advanced doctoral students; the remaining two individuals are practicing psychologists with their doctoral degree and acted as team leaders. The seven individuals were divided into two teams, each with a team leader. Each of these seven individuals was given a month of data to read individually. During this time each individual examined each message for the themes, issues, concerns, etc. of the message board participants. At the conclusion of the individual coding period, each team met to code the data together to determine whether there was consensus in their findings. Each three-to four-member team was responsible for a month (February or September) of posts. Each team used qualitative analytic tools to extrapolate themes from the data, ensuring that triangulation occurred for each theme. All of the adherence/compliance posts were then removed from the larger data set for this study.

The principal investigator then reviewed all adherence posts to confirm their relevance to the topic. It was decided that further exclusion criterion would be necessary. The additional exclusion criterion included posts about treatment adherence for other diseases/syndromes (e.g., cancer treatment) and messages concerned with pregnancy and the ethics of genetically passing long QT syndrome to children. These additional criteria resulted in 31 messages being excluded from the study.

All compliance/adherence messages were then extracted from the larger data set for in-depth study. The included messages where then reread and summarized. The principal investigator also included in the summary of posts initial thoughts and comments regarding potential themes. Messages were then read again with these initial themes in mind in an effort to determine the number of times each theme was validated
by participants’ comments. This also served as a time to redefine the initial themes that were found. For the purpose of this study, a theme was constituted by three people/messages validating the same concept in the message board. There were 68 adherence messages that were examined further for this study.

In qualitative research, it is imperative that investigators consider their own biases and the potential influence on data analysis. Throughout the process of completing the research for this study, process notes were recorded that included potential biases the research team and this principal investigator may have had about the data. The research teams involved in this larger qualitative study were all doctoral students experienced in clinical health psychology settings. Each member had familiarized himself or herself with the current LQTS research to ensure that they fully understood the user group messages.

**Types of validity.**

Qualitative research may lend itself to validity concerns because the process of achieving validity and the process of judging quality are slightly different from quantitative research. In order to ensure quality in this study, issues of validity outlined by Miles and Huberman (1994) were addressed. Miles and Huberman identify five issues of validity in qualitative research: objectivity/confirmability, reliability/dependability/auditability, internal validity/credibility/authenticity, and external validity/transferability/fittingness.

**Objectivity/confirmability.**

This type of validity is concerned with the researcher’s bias toward the data, influencing results. Studies that have objectivity/confirmability base their conclusions or
findings on the subjects’ conditions, rather than on the inquirer’s interests. This study demonstrates objectivity/confirmability through clearly described and explained methods outlining exactly how data was collected, processed, and analyzed. In addition, the principal investigator remained as self-aware as possible about personal assumptions about the data, affective states while working on the data, and considering how they may affect the results (Miles & Huberman, 1994).

*Reliability/dependability/auditability.*

These types of validity are concerned primarily with whether the process of the study was stable and consistent over time. Reliability of the process over an extended period is critical for validity. This study demonstrates reliability/dependability/auditability in its findings due to several actions taken by the researcher. For example the investigator obtained the data from a wide variety of geographic settings due to the international scope of the study. Data was triangulated, and coding checks were made by colleagues familiar with the data to ensure agreement in quality checks. The multiperson coding team also served as mechanism to check for biases and knowledge of the topic (Miles & Huberman, 1994).

*Internal validity/credibility/authenticity.*

These issues of validity deal primarily with determining if the findings of the study are logical, or make sense. Miles and Huberman (1994) indicate that there are several types of understandings that can potentially become apparent in the data. These are descriptive, interpretive, theoretical, and evaluative. To ensure that each of these types of understandings was obtained, this study used messages that were rich with context and description. In addition, while there were some areas of uncertainty (which
is to be expected), the vast majority of the messages used in the study were convincing and consistent. Rival explanations were considered throughout the research. Lastly, triangulation or consensus was found among researchers involved in the larger long QT syndrome study about the major themes found (Miles & Huberman, 1994).

*External validity/transferability/fittingness.*

This area of validity determines if study results are transferable to other contexts and populations. There are few published studies examining the psychosocial effects that a diagnosis has on individuals and families. This qualitative study can serve as the foundation for other studies (qualitative and quantitative) examining the psychosocial aspects of LQTS and may be generalizable to the population at large. Direct quotes from the sample are provided in an effort to allow potential readers to transfer experience and understanding to their own lives.
Chapter 4

Results

Data was collected from a long QT syndrome message board, online community. Messages posted between February 1, 2008, and February 28, 2008, and between September 1, 2008, and September 30, 2008, were selected for analysis for this study. Inclusion and exclusion criteria for this study related primarily to topic relevance. Messages that were related to both LQTS and treatment adherence (behavioral, lifestyle, medication, barriers, decision making, etc.) were included in analysis. Those threads that did not discuss the topic of compliance were excluded. In addition, messages related to the decision to have a child, knowing that LQTS is present in the family, were not included in this study.

A total of 711 messages were posted in the 2 months that were studied. These 711 posts were made by 89 user group participants. Of these 89 participants, 40 identified themselves as having LQTS, 41 had a child with LQTS, 10 had one or more siblings with LQTS, 4 had a spouse with LQTS, and 4 had a parent with LQTS. This analysis of data suggests that the user group members represented in this sample reflect both those with LQTS and families of persons of LQTS.

After further review, 32 posts were excluded from the final analysis because they did not meet inclusion criteria. Posts that did not reflect barriers to treatment adherence, decision making about adherence, or the challenges in compliance were excluded. Therefore, a total of 68 messages (33 from February and 35 from September) were included from 37 individuals (33 females, 4 males).
Subjects included in this subdivision of adherence posts in both the February and September data represent the global participation of this online community. Individuals came from countries including, but not limited to, South Africa, New Zealand, Scotland, the Netherlands, Vancouver, and various regions of the United. These participants also reflect the various types of LQTS diagnoses, the various treatment options, and the degree of ambiguity experienced by those with LQTS. Additional subject characteristics are provided in the table below.

Table

*Additional Subject Characteristics.*

<table>
<thead>
<tr>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Participant with LQTS</td>
<td>17</td>
</tr>
<tr>
<td>Child with LQTS</td>
<td>20</td>
</tr>
<tr>
<td>Spouse with LQTS</td>
<td>2</td>
</tr>
<tr>
<td>Sibling(s) with LQTS</td>
<td>4</td>
</tr>
<tr>
<td>Parent with LQTS</td>
<td>2</td>
</tr>
<tr>
<td>Participant/family member with current or planned ICD</td>
<td>14</td>
</tr>
<tr>
<td>Participant with AED</td>
<td>3</td>
</tr>
<tr>
<td>Participant with genetic testing completed or in progress</td>
<td>14</td>
</tr>
</tbody>
</table>
Themes.

There were two major themes revealed in the data, which were expressed through participants’ messages and conversations: loss of control and barriers to treatment. Each of these impacted compliance behavior in persons with LQTS or their caretakers. From these two overarching themes emerged several subthemes that both directly and indirectly impacted compliance behaviors in this population.

Loss of control.

The data suggests that both individuals with LQTS and their caretakers have the general feeling of having little or no control over diagnosis and treatment. One participant stated, “I think the thing that makes [long QT syndrome] even worse is not having control over what’s going on.” This sense of having no control over LQTS pervades decision making, coping, and relationships among family members. One parent stated, “I really never know what the right thing to do [is] and my wife and I fall out over it.” The lack of certainty in the diagnostic process, treatment options, and recommendations is frustrating for many people. It was well stated by a female participant, “Yes, the worst part is just not knowing.” This feeling that one has no control over long QT syndrome comes both at the time of receiving the diagnosis of long QT syndrome and throughout treatment.

This overarching feeling of loss of control was largely due to an intrinsic vagueness about both the diagnosis of LQTS and proper treatment of the syndrome. The idea that the diagnostic phase and the treatment process of LQTS were generally poorly understood by patients, caretakers, and, at times, the physicians themselves was widely evident in the data. A quote by a very concerned parent sums up these two dynamics
very well. She told the group, “Without being able to pinpoint the type of LQTS that our son has, we are left treating all the known triggers for episodes.”

*Vagueness in diagnosis.*

There were 15 occasions from the 68 messages on which patients/caretakers expressed their lack of understanding of the diagnosis. Concerns included not understanding the diagnostic procedure and qualifications (such as QT lengths), not understanding the information communicated by physicians, and confusion about disease severity. One participant expressed much frustration about the vague nature of her potential diagnosis. She states, “I, too, am confused about many of the replies that I’ve seen since I was also diagnosed with Borderline QT. My doctor says there are many considerations such as which machine was used, and other corrections so that I am close enough to borderline to check out further.” She continues at the end of her post, “Everyone’s got a different opinion so I don’t know what to think.”

There was much conversation regarding the legitimacy of borderline long QT. One participant stated in response to the confusion, “There is no such thing as Borderline long QT. You either have it or you don’t. As some people say on this group ‘you’re either pregnant or you’re not.’” Other examples of individuals feeling uncertain about their diagnosis because of the vague nature of LQTS, as well as the vague way in which medical staff introduce and explain the information, include a mother’s concern for her teenage daughter. She stated, “My daughter (15) was diagnosed with LQT5 almost 2 years ago now and ever since I have been trying to gather as much information as I could on it – I am beginning to think it doesn’t exist.” In addition, there was conversation concerning the lack of information and poor transmission of that information from the
medical community to patients with long QT syndrome. One patient stated, “I am sure that most doctors mean well but just don’t know enough about LQT to diagnose it properly.” It is this insecurity due to the vague nature of the diagnosis that increase the general feeling of loss of control among this population.

Vagueness in treatment.

In addition to a generally vague understanding of diagnostic procedures and the diagnosis itself, there is also evidence of a vague understanding of treatment protocols/recommendations. The poor education regarding proper treatment protocols for long QT syndrome seems to be the result of an inherent vagueness about proper and stringent treatment guidelines, recommendations, and restrictions. This was seen 11 times in this data set. One participant stated, “Again we have no real help or people to advise us here, we get their views but nothing to make us feel stronger or braver.” It is this lack of direction that leads to a feeling of loss of control over the diagnosis and treatment of LQTS. There were several conversations in the threads about physicians’ insecurity in diagnosis and treatment, resulting in very vague recommendations. A participant evidenced, this saying, “. . . my doctor want[ed] me to wait a month to see how things turn out before seeing a cardiologist.” She continued, “My doctor told me to keep playing recreation sports and running to keep up my fitness (1/2 marathons) as long as I don’t push myself.” It is difficult to comply with treatment recommendations that are not clearly stated or clearly understood. This repeated expression of uncertainty and vagueness about treatment recommendations and suggestions resulted in anxiety in the participants. One women stated, in her discussion of receiving the proper treatment guidelines for her new diagnosis, “I feel like I am in limbo.”
The feeling that one has no control over the diagnosis or proper treatment of long QT syndrome resulted in both noncompliant or compliant behaviors. Noncompliant behaviors were seen a multitude of times and will be further discussed in a later section on barriers to treatment. Feelings of loss of control seemed to result in three types of compliant behaviors: precompliance, hypercompliance, and pseudocompliance.

Compliant behavior, for the purpose of this study, is defined as any health-promoting behavior that has been recommended by medical doctors familiar with LQTS. This includes, but is not limited to, taking medications as prescribed, reduction in activity (depending on type of LQTS), and dietary restrictions.

Precompliance.

Compliant behavior before formal diagnosis was a repeated theme seen in this data set. On five occasions, persons mentioned engaging in compliant behavior before meeting with a specialist or before formal diagnosis had been made. Participants offered suggestions on how to act as if you had the diagnosis until a diagnosis was made. One such participant stated, “I would even suggest turning off phones, doorbells, and other startling noises under your control until you know further what needs to be done.” These suggestions would be fitting for someone with LQT2, but without a diagnosis of long QT or of the type of long QT, this may be premature. Another parent of a child with potential LQTS stated to the group, “I am trying to learn more about the best way to protect my 2 kids without prohibiting their life . . . they are active and competitive and without them having any symptoms it is hard to know what to do.” In this scenario, a mother is acting prior to the expression of any symptoms, and prior to any diagnosis.
Hypercompliance.

Hypercompliance is described as those behaviors that are compliant beyond the suggestion of physicians. There were three instances in which different families acted in this way. In one particular case, a young girl with several siblings was the only family member with long QT syndrome. The mother of this young girl told the group what she did to protect her child from harm and prevent symptoms of LQTS. She stated, “So even with the AED, we simply don’t travel . . . We sold our [vacation home] 40 min from here, as it was too far from a hospital.” She continued in the same post, “We took the money and put into a [recreation center] on our [large] property . . . We use it as a place for my husband and I to entertain while our children [very close by] if they need us to respond to [our child] . . . We do not go out for date nights.” This is an example of extreme measures being taken to prevent symptoms. In these scenarios, patients acted well beyond the normal treatment recommendations made by physicians. However, without clear guidelines or certainty for outcomes, this type of hypercompliance may be this family’s coping response or attempt to gain control over the condition and environment.

Pseudocompliance.

Pseudocompliance, or behaviors perceived as compliant or as increasing compliance, despite their irrelevance to treatment, was seen in the data on four occasions. It is likely that these behaviors were used in an effort to gain some control over the diagnosis and treatment. The following example shows how a woman with LQTS and mother of a child with LQTS believed she behaved compliantly. She stated, “. . . did a nature hike. It was a rather long (10½ miles) and the middle of summer, it was rather hot. My daughter (then 8, with LQTS), managed very well, but I suffered terribly.” Even
though I drank a lot of water, ate well, had a hat, had sun block etc. etc.” She continued, “on our next walk (2 wks later) I had good shoes, even cooler clothing, a hat, sun block, drinks, energy bars, bananas, etc., etc. . . . honest to g-d with all that preparation, what happened?”

These types of pseudocompliant behaviors also included behaviors that appeared to be spiritual in nature. A young mother stated, “I have taken my baby to a faith healer and we haven’t been back to the hospital yet, I don’t know if it was a success but I suppose that’s wishful thinking.” Pseudocompliant behavior appears to provide individuals with an added sense of control as well as a sense of comfort and safety against symptoms. However, it is clear that pseudocompliant behavior may overlap with behaviors that are against physicians’ orders.

**Barriers to treatment.**

The second major theme expressed in the data was the various barriers that patients and caretakers feel that they face in their efforts to adhere to treatment protocols. If they were given distinct and clear treatment recommendations, there were still barriers identified in the threads that can interfere with compliant behaviors. The subthemes identified were financial barriers, the decision to implant an ICD, medication adherence, and behavioral and lifestyle changes.

**Financial barriers.**

This particular subtheme was identified five times in the threads and was associated primarily with ICD placements and genetic testing. There were a few instances in which compliance was impeded by financial complications. Genetic testing is used to provide more certainty in both diagnosis and treatment. Genetic testing is quite
costly in the United States. Several participants verbalized their frustration with getting assistance for genetic testing. One woman stated, “It’s a tragedy that insurance coverage or lack of it determines if you can get these [genetic testing] done.” There was also conversation within the threads discussing the costly procedure of implanting an ICD. The decision to implant an ICD is complex. The addition of financial concerns can make the experience considerably more taxing on an individual and family. One participant stated her frustration with being able to afford the implantation of an ICD for her teenage daughter and her struggles with the insurance company. She stated, “Janet [name changed] was scheduled for her ICD placement tomorrow. Yesterday afternoon I received a phone call from the EP office saying the insurance company was refusing to give pre-authorization for it as they were determining it not medically necessary.” This financial barrier to proper treatment is not uncommon in this community.

*ICD vs. no ICD.*

The discussion about ICD placement was identified on seven occasions. Often, parents are making the decision for their child; this appeared to be the most difficult decision on the message board. One father asked the group, “Are we doing the right thing by getting him an ICD when he has been fine?” He continued, “[My son] says no way he wants it.” The decision to implant an ICD is not only a medical question, but also a question of what the child wants and his/her quality of life. There were other discussions about how a child/adolescent would feel about needing an ICD. One adult commented on how she felt an adolescent might handle the decision to implant an ICD. She offered, “I think all I could imagine was how my friends would accept me after my
arrest and after having an ICD implanted.” She continued, “Kids see the small picture, might my friends make fun of me, I will have a scar, etc.”

The decision to implant an ICD in an adolescent is also more than the medical decision; it is a decision regarding the lifestyle of that individual. Body image could interfere with the decision about ICD implantation and become a barrier to compliant behavior. There were several instances in which group members empathized with this difficult decision. One participant stated, “ICDs should be freeing, not a hindrance.” Another stated, “I realize at the age your kids are it is huge decision and they don’t want to be freaks.”

Another barrier to treatment adherence concerning ICD implantation was the belief many held that their ICD was the ultimate “safety net.” This concept, ICD as a safety net, was seen in three different threads. These individuals perceived their ICDs as insurance that allowed them to engage in behaviors that may not be considered compliant. For example, one individual clearly stated this when she said, “It’s a peace of mind thing . . . a safety-net.” The belief that an ICD can be used as the ultimate lifesaver and the tool that allows more normalcy is evident in response by a young female participant to a parent’s debate about whether or not her infant son will live a “normal life.” She stated, “I don’t know if the school will let them play in competitive sports, which I personally think is stupid if they won’t b/c the point of an ICD in my opinion is so that one can have a normal life without sudden death.” She added, “It seems they can just get an ICD and then they can do whatever they want (aside from tackle football) without worry, right?” More representation of this belief was seen in the following quote, as well: “I agreed to the betas and the ICD as a safeguard w/the intention of
continuing my active lifestyle.” This quote expresses the belief that several participants held about the protection and freedom that an ICD provides long QT patients. The implantation of an ICD, for some, is the proverbial green light to engage in more risky behaviors because the safety net is in place to provide the ultimate protection, or more accurately, the perception of protection.

Medications and their side effects

Long QT syndrome patients are usually required to adhere to a strict regimen of medications. There appear to be two ways in which medications themselves and the logistical issues surrounding medication compliance served as a barrier to proper treatment adherence. Medications and their side effects will be discussed first.

As previously mentioned, the side effects of medications are often a cause of nonadherence in many medical populations. This was also evidenced in this population of long QT patients in nine messages. In the majority of these instances, participants were concerned about experiencing negative side effects of the medication regimen. For example, one participant stated, “I believe beta-blockers have some sort of effect on my memory.” This concern has the potential to lead to noncompliant behavior and is therefore identified as a barrier to adherent behavior. In another post, a parent showed considerable concern about the LQTS medications and their negative effect on her daughter. She stated, “My daughter has to see an EP neurologist because the beta-blockers kick started migraines.” In this scenario, the use of LQTS medications was caused additional medical problems. There were also situations in which participants could not take medications. One such participant stated, “I can’t take beta-blockers. I am
intolerant to them.” In this case, his inability to remain compliant with the treatment protocol was not intentional, but a negative result related to his physiology.

*Medications, logistical issues, and drug interactions.*

Another barrier to compliance evidenced in the threads was in regard to logistical complications of taking medications as prescribed and complications and concerns regarding drug interactions. These concerns were identified 14 times within the threads. These threads were often concerned with the logistical difficulties of giving children medications. One mother stated, “Her stomach gets upset very easily without food in it before she takes her medications. So if she is unable to keep the food down she cannot take her medications. That’s when we end up back in the hospital.” This quote speaks to the concern of a mother about her daughter’s ability to remain compliant, as well as the potential ramifications of nonadherence, regardless of the intention behind the noncompliant behavior.

There were also conversations on the message board about the difficulty in keeping a child compliant with medications. In these threads, parents shared their personal barriers to compliant behavior. One mother stated. “It is hell to wake up a sleeping baby to give him his meds, we both usually end up crying.” This problem does not take place at home exclusively. In the following quote describes how medication compliance issues can extend into medical settings. This mother of a small daughter tells the group, “. . . the nurses are trying to get Mary’s [name changed] meds down her . . . such a chore that is!!!” Professional medical staff also negotiate the logistical barriers to proper medication compliance.
Another barrier to medication adherence mentioned throughout was in the context of drug interactions. These drug interactions ranged from cancer medications to over-the-counter energy drinks. There were several messages in which participants were asking for advice about over-the-counter medications, energy bars, and energy drinks. Advice such as the following was given, “yes energy drinks will certainly cause palpitations! Be careful how much caffeine you take in because that will only speed your heart.” Another example of similar advice given to the group: “Gotta watch the organic, natural and OTC [over-the-counter] items just as much as the prescriptions.” Patients and caretakers who are not made aware of this by physicians or other support staff are potentially creating risky situations in the attempt to be healthy.

Behavioral compliance: Exercise.

Possibly noncompliant behaviors, such as participating in sports and exercise, were seen in 12 messages. This type of noncompliant behavior ranged from low exertion home exercises to highly competitive exercise. There was communication in the thread regarding a significant, highly competitive sport in which a group of long QT adolescent boys participated. One mother spoke of participation by her adolescent sons’ (all of whom have ICDs) in one such competitive event. She stated, “I cried when they paddled the last race ‘cause you could see the strain in their faces and they were going for it, we had no cardiac problems, which is good, just very sore muscles.” She continued to talk about one of her sons who has long QT syndrome and his upcoming efforts. “Tom [name changed] got accepted into the Air Force provisionally, needs cardiologist clearance.” Such vigorous and competitive activity would likely be considered by most, if not all, physicians to be beyond the scope of adherence.
Behavioral compliance: Lifestyle changes.

There are also necessary changes in lifestyle with long QT syndrome, some of which may be a potential barrier to compliance. These changes include, but are not limited to, diet and nutrition, vacation, social impact (e.g., swim parties), and the ability to work outside of the home. These concerns were seen six times within the data. Often, changes in diet were necessary but difficult. A mother described the difficulty of such changes concerning her daughter’s new dietary efforts, “[The doctors] told her to drink 3x more than the average amount of fluids which is very hard to get her to do.” This necessary change would be difficult to self-regulate for an adult and is even more difficult for a mother to regulate in a child. Another example of nutritional changes being necessary and difficult was expressed by a daughter of an older LQTS patient. She explained about her mother’s recent admission to the hospital, “She lost 16 pounds of fluid before she left there and the doctor reproached her for her high salt intake.” In this quote the participant’s mother is an LQTS patient.

In addition to nutritional changes, there was also concern about children with long QT syndrome and their ability to remain compliant with treatment without interfering with their childhood. There was discussion about a children’s swim party, whether or not a child with LQTS should be able to attend, and the degree to which parents should involve themselves and remain present during such a party. In the following quote, a mother reached out to the group regarding a recent invitation her daughter received to a swim party and her concerns. She stated, “I said she could go, as long as I can go along as a lifeguard. Now she tells me the mom says no adults because she is on a tight budget and can only cater for the children.” She continues with her conflict, “Clearly the mom
does NOT know. Or thinks I am an uber-clingy mom. So . . . Lisa [name changed] reckons she can go, but no swimming (riiiight, I remember adults who tell a child ‘don’t worry, we won’t tell your mommy.’).” She ended her post expressing more concern, “Do I ruin her social life so early by banning her from swim parties?” This typifies the conflict a parent has knowing her child will most likely be nonadherent while trying to allow her daughter to live as normal a life as possible.

**Summary.**

The data set used for this study represented 37 participants in 68 messages. Each post was selected for its relevance to treatment compliance/adherence. Review of the data resulted in the discovery of two primary themes, loss of control and barriers to treatment, which were found to influence compliant behavior in this population. The overarching sense of vagueness in diagnosis and treatment of long QT syndrome, as well as the three types of compliance (precompliance, hypercompliance, and pseudocompliance) indicated, are impacted by an individual’s belief system. The health belief model can be used to understand how individual beliefs about the inherent vagueness in diagnosis and treatment of long QT syndrome, as well as the three types of compliance, impact compliant behavior in this unique population.
Chapter 5

Discussion

Long QT syndrome is an electrophysiological disorder of the heart associated with a lengthening in ventricular repolarization. Persons with LQTS present with a structurally normal heart; however, they experience rhythm disturbances associated with the lengthened repolarization. Treatment of long QT syndrome includes the use of lifestyle modifications, medications, pacemakers, internal defibrillators, and sometimes corrective surgery. The number of persons being diagnosed with LQTS is increasing in frequency due to better advocacy and diagnostic measures. LQTS affects individuals from various cultural backgrounds and all age ranges. The purpose of this study was to provide a psychosocial perceptive on this complicated syndrome and to increase understanding of compliance and the obstacles and barriers to compliant behaviors in this unique population.

There were two significant themes that emerged from the data regarding compliant and noncompliant behavior: loss of control and barriers to treatment. Each of these two overarching themes had additional subcomponents that are described in detail below.

Loss of control.

The first major theme identified in the data is loss of control. The loss of control theme represents a complex, multiple-feedback system of elements that results in either the increase or decrease of compliant behavior. The central point of this theme is the overarching vagueness inherent in the diagnosis of LQTS and the laypersons’ understanding of the condition. There appears to be vagueness in the transmission of
information from physician to patient and, in some cases, a lack of complete understanding of the diagnosis of long QT syndrome, treatment options, and proper symptom management. This vague understanding can result in distinctly different ways in which individuals understand their diagnosis and the importance of syndrome management. That is, they can interpret LQTS as potentially very dangerous or not dangerous, manageable or not manageable. The difference in these perceptions influences compliant behavior.

The data suggests that many individuals feel that diagnostic procedures and the explanation of LQTS given to them or a family member is unclear or vague. It appears that due to the multiple presentations of LQTS, coupled with the fact that there are few specialists able to provide concrete diagnoses, the diagnostic process often progresses with little explanation of the method or of the diagnosis itself. This lack of clarity about a life-changing syndrome results in people feeling that they have little or no control over the diagnosis. In addition, message board participants appeared to be uninformed of the various treatment options and various behavioral changes required in preventing LQTS symptoms. Ultimately, it seems that it is this perceived vagueness within the LQTS population that results in, for many, a feeling of loss of control, which directly impacts the decisions that lead to compliant behavior.

**Health belief model and loss of control.**

The health belief model is a psychosocial model developed to understand compliance and adherence trends in patients and can be used to understand compliance behaviors in persons with LQTS and their caretakers (Bloom-Cerkony & Hart, 1980; Janz & Becker, 1984; King, 1980). As previously mentioned, the vague nature of the
diagnostic process, the diagnosis itself, and treatment options and symptom reduction behaviors can both positively and negatively affect compliance behaviors in this population. The mediating factor is the way in which the patient or caretaker perceives the severity of the condition, the degree to which they are susceptible to the adverse outcomes (e.g., syncope, cardiac death), and the barriers to and benefits of treatment options. These are all components of the health belief model and directly impact behaviors.

People who perceive risk of and susceptibility to LQTS more highly are more likely to make the decision to act compliance with the treatment guidelines. Conversely, those who perceive less susceptibility to the adverse effect of LQTS and do not believe that the condition is serious are less likely to be compliant with treatment guidelines. In addition to these variables, perceived barriers significantly contribute to compliant behavior and will be discussed further in a later section.

In a study examining children with cystic fibrosis, three different types of noncompliant behavior were found: having inadequate knowledge of the disease, psychological resistance, and educated nonadherence (Buckingham, 1997). These themes were also found in the analysis of the LQTS data, particularly inadequate knowledge. Inadequate knowledge among many medical professionals impacts the way information regarding proper treatment is presented. Information that is not presented in a straightforward and clear manner results in a sense of vagueness. When families feel they cannot make adequate decisions about care due to vague information and treatment recommendations, it prevents them from being able to adequately estimate barriers to and
benefits of treatment. Without this information, compliance decisions cannot be made, which can lead to a feeling of loss of control and noncompliance.

*Types of compliant behavior.*

In addition to noncompliant behavior, the data analysis shows that there is a subset of compliant individuals whose behaviors indicate that there may be subtypes of compliance.

*Precompliance.*

It was evident in the data that there is a population of compliant individuals who are compliant with all treatment options and engage in all compliant behaviors to avoid symptoms before a certain or confirmed diagnosis of long QT syndrome is made. These individuals appear to perceive themselves, child, or adult loved one as highly susceptible to the dangers of LQTS and to view LQTS as a severe diagnosis. Therefore, they make the decision to comply with treatment before any definite diagnosis is given. This population will cease all activity, change their diet, and seek out any prophylactic interventions prior to any treatment restrictions formally being recommended by the physician.

*Hypercompliance.*

The data suggest that there is also a subset of individuals with LQTS who engage in compliant behaviors above and beyond the recommendations made by physicians. After diagnosis of LQTS, this subset perceives the diagnosis as very severe and is willing to implement compliant behaviors in excess in an attempt to prevent any symptoms of LQTS. This population limits travel and limits age-appropriate (physician-approved) activity, for example.
Pseudocompliance.

Pseudocompliant behavior was also seen in the analysis of data. It appears that persons engage in behaviors that they perceive as improving compliance with their overall LQTS treatment, regardless of its relationship to LQTS. This subset may also view their susceptibility as high and view LQTS as a severe condition. In addition, they regard the benefits of compliant behavior very highly. This group considers unrelated behaviors, such as wearing well-supported shoes, as increasing their overall compliance and ensuring greater safety. These pseudocompliant behaviors include superstitious behaviors.

The use of superstitious behavior in the treatment of LQTS may be a subclinical expression of obsessive compulsive disorder (OCD)-like behaviors. However, these OCD-type behaviors may be functional rather than pathological because of their protective factors. Compulsive, pseudocompliant behaviors sometimes seen in this population often served as protective factors against poor decision making and noncompliant behavior. This somewhat overcautious behavior at times appears to be OCD-like in its expression, in that the behaviors are extreme and sometimes unsubstantiated methods of preventing LQTS symptoms.

Precompliance, hypercompliance, and pseudocompliance are all types of compliant behaviors. These compliant behaviors are maintained by two related mechanisms. First, they all serve to increase patients’ and caretakers’ sense of control. These subsets of compliance create a perceived sense of control or the illusion of control over the management and unpredictability of long QT syndrome. This perceived sense of control over the diagnosis and management of the condition reduces anxiety in the patient...
and/or caretaker, thus increasing the likelihood that compliant behavior continues. This feedback system within the greater framework of compliance is one way that compliant behavior is maintained.

**Certainty in physicians’ directives.**

The data suggested that some patients and caretakers were provided with very clear diagnostic information and treatment recommendations by their physicians. When individuals in this category felt secure that the information provided was accurate and felt that the guidance they received from their medical team was correct, some patients expressed adherence to treatment, while others failed to adhere. The differences between these two outcomes seem to be mediated by dimensions of the HBM. Patients who adhere to treatment because they trust their physician’s recommendations also have a perceived sense of control over their well-being and engage in more compliant behavior. These patients and caretakers are likely to maintain more compliant behavior because there is less ambiguity and more perceived control. Patients who perceive high susceptibility to and high severity of disease and believe the benefits of engaging in compliant behaviors outweigh the barriers feel that they have more control over their health and well-being and behave more compliantly. This behavior seems to reduce anxiety and increase compliance.

Alternatively, patients who view their susceptibility as minimal risk, underestimate the severity of LQTS, and/or perceive the barriers to compliance as overwhelming are less likely to adhere to treatment recommendations. In some cases, treatment guidelines may be clearly defined by medical professionals, but the confidence to adhere to treatment is not present. In these cases, perceived barriers such as difficult
medication regimens, behavioral restrictions, or cost may be overwhelming. Despite clear information and clear treatment goals, the negative cognition of inability results in nonadherence. It is the negative cognition ("I can’t stick to this" or "This is too hard to do") that results in a feeling of incapability or loss of control, leading to noncompliance.

The perceptions of control, more specifically the perception of gaining control over this experience and diagnosis by acting in a manner that is consistent with physicians’ recommendations, reduce the anxiety provoked by the ambiguity and vagueness of the diagnosis itself. This perceived sense of control over long QT syndrome and resulting decrease in anxiety seem to increase all types of compliance (Figures 2 and 3 below).
Figure 1. Loss of control.
The second theme identified in the data is barriers to adherence. These barriers include the following subthemes: financial concerns, decisions about ICD implantation, and medication and behavioral adherence. Each of these subthemes acts as a potential barrier to adherence to the treatment of LQTS.
Financial concerns.

Patients in both the diagnosis and management phases of LQTS experience significant financial concerns. Finances appear to influence the diagnosis and management of LQTS in many ways. One area in which financial decisions impact the diagnosis of LQTS is in the area of genetic testing. There is genetic testing available for patients to determine the type of LQTS they have; however, the cost of this diagnostic tool is often prohibitive for many patients. Insurance companies may not pay for this very expensive testing. The genetic testing for LQTS assists in confirming the diagnosis, can alert individuals about other family members at risk, and directly impacts physicians’ treatment decisions. It was evident in the data that some families were unable to afford genetic testing. This inability to receive the testing and confirmation they wanted about the diagnosis seemed to result in further ambiguity about diagnosis and treatment, increasing the overarching sense of loss of control.

Other financial concerns were evident in families trying to afford ICDs and purchase AEDs for their children. The implantation of an ICD is very costly, but is generally covered by health insurance. There were situations seen in the threads in which this potentially lifesaving procedure was being questioned because of the financial burden it would cause or the complete inability to afford the device.

For many medical reasons, some children are best helped an AED rather than an ICD. There were several parents inquiring about the purchase of an AED. Discussion about purchasing an AED was concerned with the effectiveness of the device to abort arrhythmias, the financial state of the family, and deciding whether its value offsets its high cost. Often, the use of ICDs and AEDs is medically necessary to abort arrhythmias
and prevent an untimely death. However, it appears that the decision to purchase such lifesaving devices may be questioned by patients and caretakers due to financial struggles.

Financial barriers influence how individuals make decisions. This includes medical care, the cost of which can be exorbitant. When considering the health belief model, potential barriers to treatment/compliance are an important variable that can be greatly influenced by the resources of the individual or family. Financial limitations can be an overwhelming barrier to compliant behavior. Often, patients with financial barriers are, unfortunately, not given the opportunity to receive the comprehensive and protective treatments available. This type of noncompliant behavior is not intentional; rather, it is an unfortunate resultant of circumstance.

Noncompliance due to financial restriction rather than poor decision making or lack of knowledge is an area of medicine that requires advocacy. The use of health disparity research and advocacy could assist LQTS patients and their families in getting proper comprehensive care regardless of race, socioeconomic status, or geography. Mensah, Mokdad, Ford, Greenlund, and Croft (2005) conducted a study examining health disparities in a cardiovascular population in the United States. African Americans, Hispanic/Mexican Americans, those living in the southeastern portion of the country, and individuals in lower socioeconomic classes were found to have the lowest quality of life and lowest longevity.

The U.S. Department of Health and Human Services Agency for Healthcare Research and Quality produces the yearly National Healthcare Quality Reports (NHQR). With more than $2 trillion spent yearly on health care in this country, it is important to
assess the quality, effectiveness, safety, timeliness, and patient centeredness of care. Since the start of the NHQR, findings have suggested that the quality of health care in the United States continues to be suboptimal. The 2008 NHQR indicated that while there continues to be an increase in overall quality of care, there is limited improvement in the area of chronic illness management and prevention (screenings). As both a chronic illness and an illness that can benefit from prevention in early detection and intervention, long QT syndrome is one such area of medicine that requires more attention in the area of health disparity.

**ICD vs. no ICD.**

A common discussion on the LQTS online message board involved the decision whether to have an ICD implanted. One area of particular interest was the way in which the implantation of an ICD changed the compliance of some individuals, which lead to the subtheme of ICD as a safety net (Figure 3). There was a pattern seen in the threads in which patients viewed their ICD as the ultimate “safety net” and therefore engaged in noncompliant behaviors, including strenuous exercise. These individuals did not view their ICD as a last resort treatment against an arrhythmia or cardiac death. Rather, these individuals they relaxed (or ignored) the restrictions recommended by physicians and took significant behavioral risks in the belief that their ICDs would save them. In this case, ICDs were viewed as part of their treatment, rather than as the backup system if all else fails in their treatment and in the event that they experience a fatal arrhythmia. This suggests that there was a strong sense of safety and protection they felt in their ICDs, allowing them to push the envelope of compliance. In these scenarios, the ICD provided the illusion of control of their LQTS. It is likely that individuals who behaved
Decision to Implant ICD

Allows patient to be more active and do more.
- ICD as a "safety net"

Patient takes bigger risks
- "My ICD will protect me"

Patient engages in non-compliant behavior

*Figure 3.* ICD decision making.
noncompliantly due to the safety net cognition perceived a high degree of control over their LQTS care and their mortality. This perception of control or illusory control reduces anxiety and directly impacts compliance (Figure 2, above).

In addition to the concept of the ICD as a safety net, the data suggested that body image concerns also influenced the decision to implant an ICD. This mostly occurred in younger patients or parents of younger patients concerned about how the ICD would change the patient’s appearance. The implantation of an ICD results in scarring and, in some cases, a visible protrusion of the device. Concern with physical appearance after implantation impacts the decision whether to get the device. In terms of the health belief model, these individuals are likely to have decreased compliance because their perceived benefit of this surgery may be superseded by physical appearance concerns. This concern may or may not override physicians’ recommendations. Either way, it remains a concern for LQTS patients and, therefore, an area of concern for physicians and researchers.

Medications.

Medications or taking medication as prescribed is a common variable in treatment adherence. Within this long QT syndrome sample, two more specific subthemes were identified. First, many individuals expressed concern about the side effects of medications and the belief that the medications may be doing more harm than good. Rather than viewing their medications as useful and necessary, these individuals perceived their medications as harmful or uncomfortable due to some of the negative side-effects that may occur. In terms of the health belief model, negative medication side effects may be a barrier to treatment. This barrier to compliant behavior can be quite significant when an LQTS patient feels dizzy or nauseous each time he or she takes
medication. In extreme cases, patients stopped taking medications because they could not tolerate them. When a perceived barrier to treatment is so clearly experienced, as in the case of uncomfortable side effects, it negatively impacts decision making, results in feelings of loss of control, and ultimately leads to nonadherence.

The second subtheme was the logistical difficulties of medication compliance, including timing, types of medications, age of patient, and contraindicated medication. Individuals described some of the difficulties of taking medications as prescribed, especially conforming to time constraints, schedules, and other logistical issues. Young children often need to take medication during the night. Also, there were patients who, due to their age and hormonal shifts, were experiencing discomfort and had questions about compliance.

In addition, LQTS has many medications that should be avoided, resulting in confusion about what is contraindicated or the experience of additional symptoms. There was also much discussion about impact of other, more acute medical difficulties, such as the diagnosis of cancer that needed to be treated, and the impact of those treatments on proper long QT treatment. These individuals were seeking out advice on how to properly manage the treatments for two very serious medical problems.

**Behavioral adherence.**

A diagnosis of LQTS requires behavioral adherence in addition to medication and other treatment interventions. For example, strenuous and competitive exercise is not recommended for many persons with LQTS 1, and loud jarring noise should be avoided for LQTS 2. A diagnosis of any type of LQTS requires that behavioral changes be made and maintained. Such behavioral changes include (but are not limited to) exercise and
lifestyle changes such as diet and nutrition, ability to go on vacation, social impacts (e.g., swim parties), and the ability to work outside of the home. Families who highly value any one of these activities, such as competitive athletics or travel, for example, may grapple with limiting or eliminating behaviors from their lifestyles. Often, members of families identify with each other and bond over common interests and activities. A sense of loss may be felt in one family member because their LQTS treatment recommends that they not participate. This loss can be a significant barrier to treatment adherence. If a patient’s love for competitive sports or travel, for example, outweighs the degree to which they perceive their susceptibility to LQTS symptoms, it is likely that they will not comply with important treatment recommendations made by their medical team.

**Social cognitive theory and adherence.**

Health behaviors, including compliance behavior, can also be examined using Albert Bandura’s social cognitive theory. Social cognitive theory has its roots in social learning theory, a behavioral approach to understanding human behavior. Bandura (1998) discussed the application of social cognitive theory to health behaviors by primarily indicating the importance of patient (caretaker) efficacy. The perception of efficacy, according to Bandura, refers to the belief an individual has regarding their ability to plan, organize, and implement a strategy to produce a desired outcome. In this theory, self-efficacy is one of the foremost bases of change. Without the belief that one can achieve success in desired outcome, there is little desire or incentive to try (Bandura, 1998; Clark & Becker, 1998; Leahy, 2003). The belief that one can act in compliance with their LQTS treatment guidelines, despite the challenges and obstacles, may in part be mediated by the degree to which they have the confidence and desire to do so.
Efficacy can be developed and expanded in several ways. First, mastery experiences increase efficacy by providing a history of success. This history of repeated successes provides practice and quantitative evidence of ability. A second method of increasing efficacy is by vicarious experiences. Vicarious experiences are provided by trusted and experienced individuals. In this study, the message board contained many experienced individuals who have been managing their LQTS symptoms and provide models of success. Newer participants look to these individuals to provide models of and advice on symptom management and compliant behavior. For example, parents may share information on how best to administer medications to young children. The presence of reliable, trustworthy social support increases efficacy and therefore has the ability to increase compliant behavior in this population. An additional way to increase efficacy is by social persuasion. Social persuasion is used to provide positive reinforcement. Social persuasion in the LQTS message board community provided many individuals with the reinforcement they needed to believe that they have the ability to care for themselves or others, maintain health, and prevent cardiac events. Peer support decreases tension and stress, which according to Bandura and social cognitive theory results in more positive outcomes (Bandura, 1998).

Lack of control negatively affects self-efficacy and results in decreased levels of functioning and potentially poor/no treatment adherence. The pervasive theme evidenced in the data suggesting that participants feel they have little or no control of their LQTS, its diagnosis, and treatment can, according to social cognitive theory, directly impact compliance. Therefore, social cognitive theory helps to support the theory in this
research that a decreased sense of control negatively impacts compliant behavior in persons with long QT syndrome and their caregivers.

Another interesting component to this theory is outcome expectations. Individuals’ outcome expectation of both adherence and nonadherence impacts health behaviors. Positive outcomes of adherence, such as perceived symptom reduction (e.g., fewer fainting episodes), serve as an incentive to comply, while negative outcomes of adherence, such as medication side effects or cost, reduce compliant behavior. Outcome expectations are also regulated by self-sanctions (personal standards/beliefs) and serve as a mediator of health-related behaviors such as compliance. Self-sanctioned goals are rooted in individual values, which are largely influenced by efficacy. The goal for an LQTS patient to be compliant with treatment due to awareness of the risks of noncompliance is rooted in self-sanctions (values to be proactive and self-protective) and is completed by having the efficacy to try. Self-sanctions and efficacy both influence motivation behavior, which is necessary in maintaining compliant behavior for a chronic illness such as LQTS (Bandura, 1998).

**Theory of planned behavior.**

The theory of reasoned action, sometimes referred to as TRA, developed by Fishbein and Ajzen can also be used to conceptualize adherence behaviors in this population. As previously mentioned, the theory of reasoned action was developed as a method to explain the relationship between attitudes and behavior (Anderson & Lavallee, 2009; Clark & Becker, 1998; Fishbein & Ajzen, 1975; Romano & Netland, 2008). From the TRA comes the theory of planned behavior, which states that a behavior is related both to the strength of an attempted behavior and to the degree of control one has over that
behavior. Functions of control include time, knowledge, resources, support, desire, a reasonable plan, etc. (Clark & Becker, 1998). LQTS patients who feel they have little control (vagueness in information, limited financial resources, efficacy, etc.) may have a negative attitude about their ability to be compliant, which leads to a reduction in compliant behaviors. In addition, those individuals who feel that they have little or no control over treatment and diagnosis are more likely to struggle in estimating the likelihood of their ability to be successful, which negatively influences attitude and results in decreased compliance. For example, a parent with a newly diagnosed LQTS child may feel they have no control over treatment because of the vague nature of information that has been given to them by medical staff. The resulting lack of control negatively influences this parent’s ability to appropriately estimate how successful s/he will be in caring for the child. This inability to estimate success negatively influences the parent’s attitude toward his or her ability to learn about and maintain compliance, potentially resulting in fewer compliant behaviors.

In addition, the theory of planned behavior asserts that subjective norms directly impact intention. The suggestion or recommendation from a trusted or respected individual can directly influence health behaviors, including compliance behaviors. In this study, over 780 international members serve as supporters, advocates, and primarily well-intentioned friends who want to see successful outcomes in their LQTS community. The user group platform can serve as group support that can increase compliance because of the normative beliefs of compliance expressed by many veteran members and facilitators.
Limitations.

There are several limitations of this study that require discussion. First, there are limitations of the Internet user group. The message board allows many persons to participate in online conversation about a topic of interest. However, only small numbers of message board members appeared to participate regularly. These regular users may not reflect the LQTS population as a whole. Secondly, participant sampling may also not reflect the majority of LQTS persons. In addition, the criteria chosen when developing this study, primarily the decision to choose February and September entries, may not best reflect the LQTS population.

Another limitation of this study is the lack of information regarding ethnic and cultural diversity. The research team extrapolated as much ethnic, cultural, and geographic information from the messages as possible; however, this information was limited. Furthermore, the research team had limited knowledge about the intricacies of health care systems worldwide. Therefore, it is possible that information regarding health care decision making was misunderstood. Because of the global representation of the participants, there is a potential limitation in the researchers’ assumptions about the importance of psychosocial issues in health care. The research team made the assumption that the degree of integration of psychosocial considerations and health psychology issues into medical care is similar in other countries to that in the United States.

Additional limitations of this study included the ambiguity in language and context of messages used. Unlike more traditional qualitative research, which uses few subjects and lengthy interviews, this research was comprised of many participants (780 total, 89
active) participating to various degrees. Traditional qualitative interview research gives the principal investigator the ability to determine individual linguistic differences, including, emphasis, exaggeration, and emotions, for example. In addition to linguistic information, the general context of the messages could have been misunderstood due to the “snapshot” type of data that was collected.

In an effort to keep minimize limitations, the tools of qualitative research were used, including the use of questioning, the flip flop technique, waving the red flag, looking at language, looking at emotions that are expressed, “so what?”, “what if,” and looking for the negative case. Utilizing these techniques increased validity and allowed for a more in-depth and analytical view of the data. These tools are fundamental to such analysis and add to the degree to which researchers can extrapolate from the data (Corbin & Strauss, 2008). In addition to the use of these analytic tools throughout data analysis, the use of triangulation among coders also served as a method to increase validity and decrease the limitations of the study. The use of emoticons by participants also assisted with the understanding of emotional context at times.

**Suggestions for continued research.**

The data in this study was limited to those persons actively participating in the message boards during a very specific period. It is likely that if an additional longitudinal study were to be conducted, researchers may find stages or phases of compliant behavior that resemble and build upon the stages indicated in this current study (precompliance, hypercompliance, and pseudo compliance). However, this study represents a snapshot of information. The qualitative approach was taken to provide insight into the population and resulting psychological effects that surround compliance and aims to serve as a
catalyst for further research on the topic. An additional area of interest would how parental compliance influences the compliance behavior of children and those children later in life. It would serve researchers and clinicians well to understand the effects of modeling and environment on treatment adherence. Gender and its influence on compliance is another area that requires more research. There was suggestion in the data that men are less compliant than women; however, this was inconclusive. It would be beneficial to understand the influence of gender on adherence behaviors.

**Clinical implications.**

Concerns with treatment adherence in the LQTS population are warranted because of the strict compliance that is required for patient safety and longevity. LQTS patients can live long, satisfying lives and be productive members of society. However, longevity is highly correlated with the proper treatment and maintenance. Complications in treatment adherence may be prevented or reduced with proper education about LQTS and psychoeducation on adherence and its potential barriers. The inclusion of a clinical health psychologist in an integrated medical team would assist in providing education, counseling, and evaluation of potentially nonadherent patients and caretakers.

The inclusion of a clinical health psychologist into an integrated medical team such as a cardiology practice could impact patients in many ways, particularly in the area of compliance. A clinical health psychologist could evaluate new patients and help physicians identify those who may have more trouble with remaining adherent. For those at-risk patients, several steps could be taken to increase the likelihood of compliant behavior. A systematic protocol could be used to assess new and existing patients to determine the degree of intervention required. First, a thorough assessment using the
four dimensions of the health belief model should be completed. Understanding and discussing the perceived benefits of adherence would be a positive start. Next, it is important to assess whether the patient/caretaker completely understands the medical recommendations. A psychologist could ask questions such as: “What did your doctor tell you about medications?” “What do you need to take?” “How often do you need to take them?” This will help determine much the patient retained from their doctor’s appointment regarding treatment requirements and recommendations. In addition, a psychologist working LQTS patients should assess barriers to treatment. He or she could ask the patient, “What challenges do you think may arise that may make it hard to follow the doctor’s directions?” or “Can you see anything that can get in the way of taking your medicine on time, or keeping chocolate away from your child?” These questions can be used to identify potential barriers to compliance. A protocol would require assessment strategies as well as initial intervention tools that are based on components of the health belief model, social cognitive theory, and the theory of reasoned action.

The inclusion of a systematic protocol to evaluate perceptions of the condition and beliefs about vulnerability and the ability to remain in control can increase compliant behavior. In addition, these perceptions can be monitored over time. This type of care would facilitate the integrated delivery of comprehensive care in the medical setting. Munsey (2006) discussed the integration of psychology in health care, describing the way psychologists can use behavioral health understanding with the medical needs of their patients. Integrated health care is the collaboration of physicians with clinical psychologists to improve care and increase the likelihood of positive outcomes.
An important aspect and benefit of having a clinical health psychologist as a member of an interdisciplinary team of providers is assistance in the communication between caretaker/patient and physician. As seen in the messages and themes outlined in this manuscript, patients and caretakers are leaving their doctors’ visits feeling uncertain about the diagnosis of LQTS and the best way to manage LQTS. Assisting physicians in their presentation and explanation of LQTS could reduce the vagueness perceived by patients. For example, a clinical health psychologist could provide in-service education on how best to deliver bad news to families, providing additional information to help medical providers consider the developmental state of the patient, the dynamic of the family system, and the diagnostic phase of the family. The overarching sense of vagueness described by LQTS patients and their caretakers can be reduced by providing clearer information that is developmentally appropriate and delivered with unwavering clarity. This reduction in vagueness would positively influence the decision-making process, thus increasing efficacy and resulting in a potentially better treatment outcome. The collaboration between psychology and medicine would assist patients and caregivers in the long-term management of LQTS by reducing vagueness and increasing understanding of diagnosis and treatment (Munsey, 2006).

**Conclusion.**

In summary, proper LQTS management is necessary for the treatment and prevention of symptoms, including arrhythmias, syncope, and death. As explained in this paper, LQTS research is limited, and research regarding the psychosocial aspects of this illness is even sparser. This qualitative study that included individuals from around the globe offers a unique psychosocial perspective into the daily rigors of living with LQTS.
Like many other chronic illnesses, LQTS impacts the daily functioning of the patients and their families. Following the recommendations of medical staff is a necessary component of managing LQTS. This study aimed to examine the personal struggles that individuals and caretakers of those with LQTS have with treatment adherence.

There is much potential benefit that can be gained from the findings of this study. First, psychosocial research on the long QT population is sparse, and this study will add to the literature available. Second, treatment compliance in this particular population should be of great concern, as medication is necessary for the survival of LQTS patients. Third, an understanding of the psychological effects of LQTS can assist physicians in understanding their patients and the possible concerns that may arise during treatment. Lastly, this study can add to the understanding of treatment adherence not only in the LQTS population, but in any population with chronic illnesses.
References


