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Confronting the Unknown: Mothers' Reactions, Challenges, and Ways of Coping with a Child's Diagnosis of Long QT Syndrome

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CONFRONTING THE UNKNOWN:
MOTHERS' REACTIONS, CHALLENGES, AND WAYS OF COPING WITH A CHILD'S DIAGNOSIS OF LONG QT SYNDROME

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Dissertation Approval

This is to certify that the thesis presented to us by Twain Gonzales on the 17th day of July, 2009, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

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DEDICATION

This dissertation is dedicated to the mothers who openly and warmly volunteered to participate in this study. It is their courageousness, honesty, and love for their children that brought this research to life. It is their stories which will assist professionals in obtaining greater understanding and skills in order to provide more effective services to those living with long QT syndrome.

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ABSTRACT

Long QT syndrome (LQTS) is a life-threatening inherited cardiac condition. It is caused by a disturbance in the ion channel genes that control cardiac repolarization, resulting in ventricular arrhythmias that can lead to syncope, sudden cardiac arrest (SCA), or sudden cardiac death (SCD). Most deaths occur in children, adolescents, and young adults. Although congenital LQTS occurs in 1/3000 to 1/5000 individuals, to date, little research has been conducted on the psychosocial effects of LQTS. Mothers who are confronted with a new diagnosis of their children’s LQTS are likely to have never heard of the disease prior to diagnosis, and few psychosocial resources may be available. The aim of this research was to conduct a qualitative study that focuses on mothers’ coping abilities regarding the uncertainty of LQTS and how their quality of life is affected on a daily basis. Ten mothers were randomly selected and interviewed in a semistructured format. Each interview was audio taped and analyzed using a grounded theory method.

The primary objective of this study was to acquire descriptive data through qualitative analysis to assist mental health professionals and health care providers to (a) identify how mothers cope with the diagnosis of LQTS, (b) determine the effects of LQTS on the quality of life, (c) understand the role of uncertainty and unpredictability of LQTS in daily coping, (d) parenting challenges, (e) and identify effective problem-solving.

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CHAPTER 1

Introduction

Background of the Problem

Long QT syndrome (LQTS) is an inherited or mutated genetic defect within the muscle cell structure of the heart, namely, the ion channels (Moss, 2009; QTsyndrome.ch, 2006). Because of the abnormality of the firing of the ion channels, an inappropriate electrical charge is generated; as a result, the electrical system of the heart becomes irregular, thereby generating an arrhythmia. Specifically with LQTS, the natural rhythm of the heart is prolonged during an interval known as the QT interval, which is caused by an overload of myocardial cells with positively charged ions during ventricular repolarization (Goldenberg & Moss, 2008; Moss, 2009; QTsyndrome.ch, 2006). The prolonged QT interval predisposes individuals to a rapid heart rhythm known as torsade de pointes (TdP). Unfortunately, TdP can result in syncope and cardiac arrest (Ashworth, Levsky, Marley, & Kang, 2005; QTsyndrome.ch, 2006). Although the actual prevalence of LQTS is unknown, it is suspected to be a common cause of sudden and inexplicable death in children and youth (QTsyndrome.ch, 2006).

Currently, it is estimated that 50,000 individuals in the United States are affected with LQTS, resulting in 3,000 deaths annually (QTsyndrome.ch, 2006). In addition, it is projected that 1 in 3,000 to 1 in 5,000 infants are born with LQTS (Goldenberg & Moss, 2008; Moss, 2009). In fact, in 1998 the *New England Journal of Medicine* reported that half of the infants who died of sudden infant death syndrome (SIDS) in Italy were also afflicted with LQTS; 7 of every 10,000 newborns die of SIDS in Italy annually (QTsyndrome.ch, 2006).

In the United States, the Mayo Clinic suggests that approximately 5% of SIDS is due to LQTS (QTsyndrome.ch, 2006). Although SIDS is the leading cause of death among infants between 1 month and 1 year of age and strikes 1 or 2 of every 1,000 babies, no theory regarding the etiology of SIDS has been fully supported (QTsyndrome.ch, 2006). For example, if the infant dies of a ventricular fibrillation during the first few months without an electrocardiogram (EKG) screening, the cause of death would likely be mistakenly attributed to SIDS (QTsyndrome.ch, 2006). Therefore, the full effects of LQTS on SIDS are unknown at this time, likely resulting in underreporting of its prevalence.

LQTS as a Chronic Illness

Estimates suggest that 30% (Wise, 2007) of children under the age of 18 have a chronic illness; approximately 6.5% of these children have a disabling chronic condition (Tak & McCubbin, 2002). Although considerable research exists regarding parents' general reactions and adaptation to their children's chronic illnesses, specificity regarding parental response and adaptation to the onset of a chronic illness (Hauenstein, 1990) in addition to other factors is lacking. For example, literature suggests that parents' adaptation and adjustment may vary depending upon the nature and course of the chronic condition (Dodgson, Garwick, Blozis, Patterson, Bennett, et al., 2000), their knowledge of the condition, and the suddenness of its onset. In addition, parents' problem-solving abilities and problem orientation (Chang & D’Zurilla, 1996) may influence their adaptability regarding how to cope with the onset of a child’s chronic illness. Unlike other chronic conditions, the diagnosis of LQTS is relatively new, and it is a chronic illness for which the onset, course, and prognosis are uncertain.

For those in whom LQTS is diagnosed before any serious symptoms appear, current treatments grant them the possibility of a normal lifespan, depending on the genotype of the condition. However, contending with a chronic illness that has no specific remedy, an uncertain prognosis, and no guarantee of an uninterrupted or full lifespan can leave both the individual who has LQTS and the individual’s family with confusion and distress (Brown, 1993; Cohen, 1993a; Cohen, 1993b; Dodgson, Garwick, Blozis, Patterson, Bennett, et al., 2000; Hauenstein, 1990).

Unlike other chronic illnesses, which have a clearly defined progression of the disease and/or clearly defined symptoms, LQTS is a chronic condition with no obvious symptoms until a serious incident occurs, such as syncope, torsade de pointes (TdP), sudden cardiac arrest (SCA), or sudden cardiac death (SCD) (QTsyndrome.ch., 2009). Other than genetic testing, screening via electrocardiograms remains nonconclusive and is only an indicator. Unfortunately, because LQTS is understudied and is not in the public consciousness as a serious and prevalent threat, insurance is only beginning to cover the cost of genetic testing in certain cases. Therefore, those families who lack financial resources generally opt out of the genetic testing, leaving family members at risk and without proper intervention and treatment. Estimates suggest that the cost of genetic testing for LQTS is approximately $5,400 for the individual afflicted with LQTS and

$900 for each additional family member (Cardiac Arrhythmias Research and Education Foundation, 2007).

The importance of genetic testing lies in the various genotypes of the syndrome. Currently, 12 known genotypes exist, as well as 800 mutations with significant clinical differences among the first three genotypes (Moss, 2009). The first three genotypes are LQT1, LQT2, and LQT3, each of which carries different risk factors and environmental triggers. For example, genotype LQT1 is triggered by exposure to strenuous exercise, such as running and/or swimming, and can precipitate a fatal cardiac incident (Moss, 2009, QTsyndrome.ch, 2006). In fact, concerns have arisen that some drowning deaths may actually be the result of an LQT1 incident, thereby contributing to the underestimation of the mortality and appropriate diagnosis of LQTS. Similarly, LQT2 is known to be triggered by emotional stress and startling noises, such as an alarm clock (Moss, 2009, QTsyndrome.ch, 2006). Finally, an infant or child with LQT3 may be at risk while sleeping (Moss, 2009, QTsyndrome.ch, 2006), leading to SCD or misdiagnosed SIDS.

Although beta-blockers and/or the implantation of defibrillator devices into the child's body may assist in stabilizing particular genotypes of LQTS, parents are still confronted with the unknown of their child's physical health and well-being in the present as well as the future. The uncertainty of this particular chronic condition and the lack of current psychosocial research sets this chronic illness apart from others. In fact, an extensive review of literature indicates currently only two published studies related to LQTS and its psychosocial effects on the patient or the patient's family members (Farnsworth, Fosyth, Haglund, & Ackerman, 2006; Hendriks, Grosfeld, van Tintelen, van

Langen, Wilde, et al., 2005). Other research to date is medical in nature. Literature on parents of children with asthma, diabetes, and other chronic illnesses provides some guidance on what might be relevant considerations for this population. However, even these chronic illnesses lack research related to the effects of the uncertainty of the illness upon the family structure (Dodgson et al., 2000; Jessop & Stein, 1985; Patterson & Garwick, 1994; Rolland, 1994).

Statement of the Problem

Mothers dealing with less known chronic conditions, such as LQTS, are confronted with distinctive challenges as compared to chronic conditions that have a specific course of progression and/or outcome. For example, mothers coping with such chronic conditions as asthma or diabetes do not have the threat of SCA or SCD. Therefore, mothers of children in whom LQTS has been newly diagnosed contend with a greater sense of uncertainty about the condition and its potential lethal consequences. This likely leads to a heightened sense of the mothers' perception of their children's vulnerability knowing that their children could go into SCA or, even worse, SCD. The perception of children's vulnerability as well as mothers' own coping resources will impact both mothers' and families' quality of life (QOL), as well as their own psychological and emotional adjustment.

Purpose of the Study

The purpose of this study was to investigate how mothers of children with LQTS react and adapt to a chronic condition for which no definitive outcome exists, as well as how the variables of perceived vulnerability and uncertainty impact maternal coping. Utilizing the grounded theory method, the mothers of children with LQTS were asked to describe what it was like to cope with the illness, how it affected their quality of life, and parenting challenges unique to LQTS. Additionally, various coping and problem-solving styles in contending with the disease were examined.

Definition of Terms

Automated external defibrillator (AED): A device that automatically monitors the heart rhythm and, if it detects a problem, delivers an electrical shock to restore a normal heart rhythm (Mayo Clinic, 2006).

Chronic illness: A disease that is long lasting or recurrent. The term *chronic* describes the course of the disease, or its rate of onset and development (Brown, 2003).

Coping: The process of managing taxing circumstances, expending effort to solve personal and interpersonal problems, and seeking to master, minimize, reduce, or tolerate stress or conflict (D’Zurilla & Chang, 1995).

Diagnosis: The identification of the nature of anything, either by process of elimination or other analytical methods (Merriam-Webster Medical Dictionary, 2009).

Electrophysiologist (EP): A specialist in electrophysiology (Merriam-Webster Medical Dictionary, 2009).

Electrophysiology: A study of the electrical phenomena that is associated with the physiology of the body (Merriam-Webster Medical Dictionary, 2009).

Etiology: The study of why things occur, or even the reasons behind the way that things act, and is used in philosophy, physics, psychology, government, medicine, theology and biology in reference to the causes of various phenomena (Merriam-Webster Medical Dictionary, 2009).

Implantable cardioverter defibrillator (ICD): A small, battery powered electrical impulse device which is implanted in patients who are at risk of sudden cardiac death. The device is programmed to detect arrhythmia and correct it by delivering an electrical charge to the heart (Mayo Clinic, 2006).

Prognosis: A medical term denoting the doctor‘s prediction of how a patient will progress and whether there is a chance of recovery. This word is often used in medical reports of a physician’s view on a case (Merriam-Webster Medical Dictionary, 2009).

Quality of life: Used in health care to refer to an individual’s emotional, social, and physical well-being, including their ability to accomplish the ordinary tasks of living. It may also include a person’s perception of his or her position in life within the context of his or her particular cultural milieu, as well as of maintaining a value system in relation to his or her goals, expectations, standards, and concerns (American Thoracic Society, 2007).

Sudden infant death syndrome (*SIDS):* A syndrome marked by the symptoms of sudden and unexplained death of an apparently healthy infant aged 1 month to 1 year (Merriam-Webster Medical Dictionary, 2009).

Syncope: Loss of consciousness resulting from insufficient blood flow to the brain (Merriam-Webster Medical Dictionary, 2009).

Torsade de pointes (TdP): Ventricular tachycardia that is characterized by fluctuation of the QRS complexes around the electrocardiographic baseline and is typically caused by a long QT interval (QTsyndrome.ch., 2006).

CHAPTER 2

Literature Review

The purpose of this chapter is to review published articles pertaining to the experiences of mothers and fathers of children with chronic illnesses. The literature also provides information on the parents' challenges of coping and surviving with their children's disease. The review of literature further provides studies that discuss the psychosocial impacts on this population of parents and provides data on the emotional effects on the parents with a child with chronic illnesses. The review of literature will help to build the current body of knowledge about how LQTS originates and its effects on the family's quality of life.

Theoretical Underpinnings

Parental Response to a Child's Chronic Illness

Parental reaction to their children's diagnosed chronic illness. Brown (2003) conducted a qualitative grounded theory study exploring parents' experiences in having a child with congenital heart disease and reported the effects of an initial diagnosis of a child's chronic condition upon parents. This is often described as overwhelming. In addition, the parents in her study reported feelings of shock, guilt, frustration, exhaustion, anxiety, hopelessness, fear, anger, and lack of control. Parents also expressed feeling inadequate to function in their role as a parent. In addition, during the initial reporting of the condition by the medical establishment, especially related to the initial hospital visit, parents reported distress and helplessness by stating that they felt unable to protect their child from the illness and found it daunting to make decisions for their child under the given circumstances.

Brown (2003) also found that the rapid onset of the disease or its unexpectedness contributed to increased stress among parents first hearing the diagnoses. In addition, parents' knowledge about the illness and its potential effects are likely to contribute to their stress reaction. Parents of children with congenital heart disease and other chronic illnesses reported increased stress associated with their children's expressions of fear or sadness as well as with children's inability to communicate during the initial hospitalization (Board & Ryan-Wenger, 2000; Brown, 2003).

Cohen (1993a) describes parents' experience upon hearing the news of their children's chronic and life threatening illness of cancer as shattering the parents' prior reality. Simultaneously, the parent must rapidly assimilate large quantities of highly complex medical information, generally conveyed in ambiguous language, regarding prognosis and outcome. As a result, parents who had little reason to question their parental roles are confronted with significant questioning about their roles as parents (Cohen, 1993b). In fact, Board and Ryan-Wenger (2000), who conducted a meta-analysis of 38 studies of the experiences of parents whose children were admitted to pediatric intensive care units, suggest that perceived changes in the parental role due to the newly diagnosed chronic illness may be reported as one of the most stressful aspects of adjustment. Unlike children with LQTS, most children with other life-threatening or chronic illnesses, such as congenital heart disease and cancer, generally have symptoms, and parents are able to measure the progression and treatment of their children’s disease. Although an uncertainty may remain about the effects of the treatment, specific diagnostic benchmarks exist that can eventually assist parents in knowing the eventual outcome of the illness. Those parents with children who have LQTS have no such benchmark, only uncertainty as to if and when the chronic condition could take its toll. Additional studies of parents contending with other chronic illnesses, such as children in whom type 1 diabetes has been newly diagnosed, report that hearing and assimilating the new diagnosis of the chronic condition of diabetes in their children may result in symptoms of posttraumatic stress for the parents (Landolt, Ribi, Laimbacher, Vollrath, Gnehm, et al., 2002).

Landolt et al. (2002) suggest that parents’ perception that their child is threatened with the possibility of premature death generates the beginning symptoms of posttraumatic stress disorder (PTSD). This perception, along with how the parents cope with the news, determines whether the parents develop PTSD. The age of the child, the child’s reaction, and the types of medical testing and intervention that needed to be performed contribute to parents’ stress factors. In support of PTSD symptomatology, Chaney et al. (1997) and Thernlund, Dahlquist, Ivarsson, Ludvigsson, Sjoblad, and Hafflof (1996) suggest that during the first several months after learning of their children’s diabetes, those parents demonstrate significant signs of moderate to high distress, resulting in the possibility of impairment in parental functioning.

Once again, it must be stated that common elements exist among parents of children with type 1 diabetes and parents of children with LQTS regarding uncertainty about their children’s condition. However, diabetes is generally a treatable disease, with clear measures and benchmarks as to the children’s condition on a daily basis. For example, a daily glucose test can determine and assist parents in regulating their children’s insulin levels. However, parents with children contending with LQTS face the uncertainty and lack of measurable outcome of the children’s current medical status throughout the lifetime of the children.

Parents’ reactions to the unpredictability of LQTS. Parents of young children who have a chronic condition that lacks predictability of symptoms and course of treatment are more likely to exhibit higher distress (Dodgson et al., 2000). Unlike childhood diabetes, which has measurable blood glucose levels that allows the parents some ability to monitor the nature and course of the disease, LQTS is an erratic and immeasurable chronic condition. Patients with LQTS can live a normal lifespan with no evidence of symptoms, while others may have a one-time event that can be fatal. Some individuals fall in between the two extremes. Although psychosocial research has been conducted with adults living with an unpredictable chronic condition, relatively little research has been conducted with children and the effects of chronic disease upon their parents. Unpredictability and uncertainty of the condition also affect parents’ stress levels and perceptions of their children’s condition. Dodgson et al. (2000) suggest that, for those conditions for which no predictable course or symptoms exist, parents need to be taught how to cope with and manage the ambiguous nature of the illness.

Diverse chronic conditions and differential parental reactions. Currently, little research has attempted to systematically distinguish the impact of type of childhood chronic illness upon parents’ and/or families’ adjustment levels (Patterson & Garwick, 1994; Tak & McCubbin, 2002). The few studies which have been conducted demonstrate mixed results. For example, Holden, Chmielewski, Nelson, and Kager (1997) found “family stress variables and disease-related factors that can be conceptualized across chronic illnesses such as asthma and diabetes are more salient predictors of adjustment outcomes than individual illness categories” (p. 24). In contrast, Goldberg, Morris, Simmons, Fowler, and Levison (1989) found that parents of children with chronic heart disease exhibited higher stress than parents of infants with cystic fibrosis. In contrast, “parents in the cystic fibrosis group reported more stress arising from child demandingness” (p. 355). This suggests that disease type and other factors such as onset, prognosis, and nature of illness all affect family and parental functioning. In addition, Tak and McCubbin (2002) state that different chronic conditions exhibit different onsets, varying outcomes, and an array of symptoms and courses of treatment, leading to varying stress reactions from families. Hauenstein (1990) contends that although children from different chronic condition categories may have some common experiences, the parental distress level is associated with additional perceptions of the chronic illness type itself. Depending on the severity of the illness, along with the type of caregiving responsibilities (Hauenstein, 1990) associated with a particular condition, different stressors and demands are thereby placed upon the parents. In addition, other research suggests that parents’ perception of the severity of the illness affects the parents differently (Board & Ryan-Wenger, 2000), promoting the concept that different chronic conditions will generate different parental stressors.

Another consideration regarding various childhood chronic conditions and their different effects on parents’ reactions is newer and less well-known conditions, such as LQTS. One may speculate that a newer disease for which a lack of public awareness exists, as well as vague information about the condition itself, may possibly generate a greater level of uncertainty and distress. If this speculation holds true, parents of children with LQTS will likely exhibit different perceptions and reaction formations than parents of children with other types of chronic illnesses. Again, to date, the only information from which to extrapolate how parents contend with LQTS must be based on the parents' experience, by and large, with other chronic illnesses.

Factors Influencing Parents' Ability to Cope With a Child's Chronic Condition

Perceived vulnerability. For the purposes of this study, perception is defined as an individual's cognitive appraisal of a situation or experience. With regard to cognitive appraisal, Oliver and Bough (2002) states that "cognitive appraisal is central to stress and coping, for it determines how an event is perceived and therefore operates as an essential mediator between the event and the outcome" (p. 2). Lazarus (1991) expands on the definition of cognitive appraisal and its relation to coping by suggesting that it is an individual's subjective or abstract evaluation (appraisal) of the event occurring in the external world that will determine a person's sense of well-being or stress. Finally, Oliver and Bough (2002), in a review of literature, suggests that depending on how one interprets or attributes meaning to a particular event, rather than the impact of the actual stressor, will determine the psychological impact upon one's well-being.

Given this definition, parents' perception of their children's vulnerability is a highly disconcerting factor in coping with chronic illness. Perceived vulnerability generates a sense of uncertainty about the children's welfare and also may call into question one’s perception of self as a competent parent or caretaker. Brown (2003) conducted a qualitative study of 37 parents of children with congenital heart disease. She discovered that those parents began to call into question their own parental competence. In addition, Brown (2000) discovered parents’ sense of personal inadequacy intensified when they were required to learn and manage the daily medical and caretaking regimen of their child. Deford (1983), in his book, describes the personal assault upon his self-perception and competence as a parent when learning of his daughter’s diagnosis of cystic fibrosis. He depicts his emotional and psychological reaction as contradictory in nature. He explains this concept by perceiving himself as both the creator of his child’s life, yet simultaneously feeling responsible for the child’s pain and pending death, leading to a sense of incompetence and inadequacy (Deford, 1983). Consequently, the more vulnerable a parent perceives their children’s well-being to be, the more uncertainty about the children’s welfare is likely to be generated. Forsyth, Horwitz, Leventhal, Burger, and Leaf (1996) support this notion by suggesting that the more a parent’s perception of their child’s vulnerability increases, the more likely cognitions of anxiety regarding their child’s susceptibility and mortality are to be spawned. In fact, Green and Solnit (1964) coined the term *vulnerable child syndrome* to explain the phenomenon of pathological anxiety that can occur on the part of the parent who ruminates over the possibility of the loss of their child from an illness or chronic condition (p. 58). In short, the vulnerable child syndrome is a descriptor explaining the maladaptive pattern of interaction between the parent of the ill child and the child himself or herself. This maladaptive perception and interactional pattern can foster undue psychological distress in the child, leading to psychosocial and developmental difficulties for both the parent and the child (Forsyth, Horwitz, Leventhal, Burger, & Leaf, 1996; Green & Solnit, 1964).

Youngblut and Shiao (1992) conducted a study of parents of young children under the age of 5 who were admitted to a pediatric intensive care unit for a variety of illnesses, such as congenital heart disease, epiglottitis, neurological diagnoses, and a variety of operable conditions, to determine parents' reactions and perceptions to the severity of their child's illness. They report that the parent's perception of their child's illness is not objective in proportion to the severity of the condition. In fact, the study goes on to suggest that as the result of parents' subjective perception, greater distress for the child's future may develop, along with more confusion associated with their role as a parent. Therefore, parents' perceptions of their child's well-being will likely have a significant impact upon their ability to cope effectively with more elusive chronic conditions such as LQTS.

Understanding and knowledge of illness. Lack of knowledge of a particular illness is another factor that may influence parents' perception of the chronic condition. For example, Dodgson et al. (2000) conducted a 5-year longitudinal study with 173 mothers and 150 fathers of young children with an array of chronic conditions accompanied by unpredictable symptoms. The chronic illnesses included congenital heart disease, cerebral palsy, spina bifida, cystic fibrosis, and Down's syndrome. The comparison group consisted of parents of children with predictable symptoms but unspecified illnesses. They found both fathers and mothers of children who had unpredictable symptoms exhibited higher distress in contrast to parents of children with predictable symptoms. Their conclusion suggests that parents who have a greater understanding, knowledge, and ability to predict the illness will exhibit less distress regarding their children's medical condition.

Etiology of illness. Cohen (1993a) examined the etiological uncertainty experienced by parents of children with cancer, hemophilia, congenital heart disease, cystic fibrosis, and Lowe's syndrome. She found that parents who either had the illness themselves or other family members who had the same or similar disease as their child did not exhibit the same profound need for an explanation of the etiology, whereas for parents who are "first-generation" (meaning the first time the condition is exhibited as a phenotype), even if a well-established and unambiguous explanation of the inheritance of the condition is demonstrated, the explanation of the etiology may still not be satisfactory for some of the parents. Cohen (1993a) describes that parents who are highly distressed and do not have previous experience with their child's disease tend to search for causal and even irrational cognitive links between the condition and the etiology. For example, she reported that in order to cope with the etiological uncertainty, many "first generation" parents will often create their own cognitive scenarios or distortions regarding the genesis of the condition, such as it resulting from God's retribution for their ". . . wrongdoing, a test of faith, exposure to radiation" or any other rationalization to serve as a causal link (Cohen, 1993a, p. 141). She continues by suggesting that the absence of a cognitively satisfying causal link (even if distorted) leaves the highly anxious parent feeling defenseless, with a lack of control. In Cohen's words, the parent would rather feel a sense of control over the illness by believing they did something wrong than to believe they live in a "chaotic and precarious world of unpredictable and uncontrollable forces" (Cohen, 1993a, p. 141).

Cognitive distortions. Cognitive distortions are errors in thinking (cognitive processes) and the misinterpretation of events (Nezu, Nezu, & Lombardo, 2004). Freeman, Pretzer, Fleming, and Simon (2004) refer to cognitive distortions as systematic and dysfunctional thinking processes and patterns which contribute to misperceptions of social and interpersonal interactions. Beck (2008) proposes that patterns of negative thinking generate one's distortion or perception of reality. According to Beck, negative patterns or cognitive distortions lead to depression and other pathology. In addition, Beck also suggests that negative cognitions may have been formed early in one's developmental history (Beck, 2008). Thus, negative cognitions or distortions are likely to affect how one copes with and interprets current stressors and events unless otherwise modified.

Upon understanding the role of cognitive distortions, it may be surmised that each parent's particular cognitive distortion will have a considerable impact upon their perception of the children's chronic illness. According to research on parents of children with chronic illnesses, distortions may include concerns of being punished for a wrongdoing, while others may think it has something to do with their age or that it is a test of faith sent by God (Cohen, 1993a). Deford (1983) discusses the concept that some parents may contend with the cognitive dissonance of believing they gave life to their children and are now responsible for the children's pain, actually seeing themselves as the torturer. Several studies found that parents' appraisals of the disease or chronic condition will affect their mental health and level of distress (Brown, 2003; Cohen, 1993a; Deford, 1983; Ireys & Silver, 1996; Tak & McCubbin, 2002). For example, some parents report feeling guilty for their children's disease, even though the parents had no control over the illness (Brown, 2003; Dodgson et al., 2000).

Chronic conditions studied to date relate to the more familiar illnesses for which clearer medical directives and information on the course of the disease exist, rather than the lesser known conditions. This does not diminish or invalidate the distress and trauma of parents of children with more familiar conditions; however, it is proposed herein that parents who have children with LQTS may have a great sense of uncertainty, high perception of child vulnerability, and a variety of other factors affecting their distress and coping ability in relation to LQTS. Therefore, parents whose children have LQTS may be at higher risk of distress than or exhibit different reactions from those parents whose children have a well-known disease with a specific prognosis and predictable symptoms.

Chronic Illness and Quality of Life

Quality of life is defined as an individual's satisfaction with various aspects of his or her life that he or she deems important (American Thoracic Society, 2007). The American Thoracic Society (2007) continues the definition by suggesting that a person's quality of life incorporates a concept of well-being or wellness in a variety of domains, including but not limited to physical, mental, emotional, social, and spiritual health. Quality of life may also include a person's perception of his or her position in life within the context of his or her particular cultural milieu, as well as of maintaining a value system in relation to his or her "goals, expectations, standards and concerns" (World Health Organization Quality of Life Group, 1997). Clearly, as parents attempt to adjust to the news of their children's LQTS, quality of life will be impacted. For example, adjusting to the uncertainty of the illness, and contending with the lifelong daily threat of their children's death due to LQTS and the unpredictable nature of the illness, leaves parents questioning and adjusting to a new perception regarding the quality of their lives and their children's lives.

Coping With the Uncertainty of LQTS

Defining coping. Lazarus (1999) describes coping as a set of cognitive and behavioral actions aimed at managing difficulties and stressors in everyday living, as well as the affective component related to those difficulties. McCubbin and McCubbin (1993) explain coping as specific cognitive and behavioral processes by which a person or family attempts to reduce and/or manage demands in the face of a health crisis. Coping with the uncertainty and ambiguous nature of chronic conditions such as LQTS has been identified as a significant stressor that influences family adaptability (Dodgson et al., 2000). Uncertainty about a child's LQTS is one such challenge or demand for which appropriate and effective means of coping must be employed if parents are to function effectively in their milieu while simultaneously providing a nurturing and supportive atmosphere for their child. Although some parents of children with LQTS may adapt well to their child's chronic condition, others may find coping with the uncertainty of a condition distressful, thereby leading to psychosocial difficulties and maladjustment.

Coping with uncertainty and chronic illness. Although significant research has been conducted on effective coping strategies related to daily stressors and a variety of medical illnesses, relatively little research has been explored in relation to coping with uncertainty as it pertains to chronic medical conditions (Dodgson et al., 2000; Jessop &

Stein, 1985; Knafl, Breitmayer, Gallo, & Zoeller, 1996; Rolland, 1994). Dodgson et al. suggest that research is needed to determine if families and parents of children with chronic conditions with unpredictable symptoms and uncertainty regarding the course of the illness and/or uncertain life expectancy are at greater risk for distress.

Empirical research on coping with uncertainty and chronic illness. The relatively few studies that have been conducted on parents' coping with distress regarding their children's chronic illness and uncertainty suggest that families' attempts at managing uncertainty are similar across diagnostic categories when it comes to children and the unpredictability of chronic conditions (Cohen, 1993b; Hauenstein, 1990). For example, Cohen (1995) found that parents coped on an informational basis; in other words, parents tended to be so overwhelmed with the uncertainty that they were only able to process a limited amount of information regarding the condition. Parents become selective listeners and are initially only able to comprehend rudimentary elements of the information presented to them. Over time, parents slowly begin to assimilate the actual ramifications of the condition and continue to cope on a day by day basis.

In their study of 323 parents of children with chronic illnesses, Dodgson et. al. (2000) found that families whose children's chronic condition was accompanied by unpredictable symptoms were generally more overwhelmed and distressed than those families whose children had a chronic illness with predictable symptoms and outcomes. They also suggest that a child's developmental needs also act as "trigger points," which may increase the parent's sense of uncertainty as a competent caretaker (Dodgson et al., 2000). In addition, Dodgson and colleagues (2000) suggest that parents of children with unpredictable symptoms may be better able to cope if they are provided the opportunity to engage in ongoing dialogue with professionals so they may process the information on a consistent and routine basis related to the time frame that fits both the family's and child's needs. Finally, they recommend that discussing parents' concerns regarding the unpredictability of the child's symptoms will aid the family and improve the parents' ability to cope with the ambiguous and uncertain nature of childhood chronic conditions (Dodgson et al., 2000).

Coping and other medical disorders. With regards to coping and other medical disorders, a variety of coping strategies are effective. Brown (2003) conducted a qualitative (grounded theory) study with parents of children with congenital heart disease and discovered five primary themes of coping that assisted parents when first learning about their children's condition: support and availability of family and friends, access to the child during medical hospitalization and treatment, spirituality, talking with parents going through a similar experience, and the support of medical staff who exhibited approachability. Significant research exists suggesting social support, family cohesiveness, and the family's relationship to health care workers all support positive coping in the face of medical illnesses. Other studies have found that individuals relying on such factors as hope, optimism, humor, and religiosity while contending with health in general as well as such chronic illnesses as cancer and amyotrophic lateral sclerosis (ALS) are also better able to cope (Bremer, Simone, Walsh, Simmons, & Felgoise, 2004; Lefcourt, 2005; Pargament & Mahoney, 2005; Peterson & DeAvila, 1995).

Ineffective coping. The inverse is also demonstrated to be true; those who are unable to cope effectively in the face of uncertainty of chronic illness, such as LQTS, are likely to exhibit symptoms of depression, anxiety, and other symptomatology of maladjustment. For example, Nezu and colleagues (1999) found that depressive disorders developed as the consequence of ineffectual social problem solving (a form of coping). According to Chang and D'Zurilla (1996), without the appropriate coping and problem-solving ability, psychological distress, maladaptive coping mechanisms, and an impaired sense of psychological well-being are likely to be experienced. This can be seen in such maladaptive coping mechanisms as alcohol and substance abuse, denial and avoidance behaviors, and social isolation. Therefore, constructs of effective coping with parents of children with chronic conditions for which uncertainty remains a distressful factor need to be addressed. As demonstrated by the aforementioned research, parental perceptions such as hope, optimism, spirituality, and humor, as well as proximity to the ailing child, knowledge about the condition, and social and professional support all contribute to adaptive coping. However, because such factors are somewhat vague, it is suggested a more empirical model of coping be utilized as a guide to the exploration of the constructs associated with coping and the uncertainty of the less known chronic illness of LQTS.

Social Problem Solving and Coping With LQTS

Social problem solving defined. Social problem solving is defined by Nezu et al. (1999) as a metacognitive process by which individuals attempt to comprehend the nature of their problems in managing their daily lives while directing their efforts at coping with the difficult situations. Social problem solving is demonstrated by the manner in which individuals process information about their self, the environment, and the problems encountered in everyday life, including a new diagnosis of a chronic illness. D'Zurilla and Nezu (2007) define social problem solving as the cognitive-behavioral process by which an individual attempts to identify stressors in everyday living and attempts to implement effective and adaptive solutions.

Just as with the concept of general coping processes described in the previous section, not all attempts at social problem solving are necessarily effective. Research in social problem solving has demonstrated that individuals display several problem-solving styles (D’Zurilla & Nezu, 2007). The specific problem-solving style, and the skills that lie therein, will determine the adaptiveness and effectiveness of the individual’s attempt to cope with the problem or crisis at hand. For example, the utilization of the wrong or ineffective social problem-solving style leads to increased frustration, stress, negative mood, and maladaptive psychological and physical states (Nezu, Nezu, Friedman, Faddis, & Houts, 1998), whereas an individual who has learned to employ more adequate social problem-solving skills will likely decrease and modify the distress, anxiety, and other psychosocial consequences associated with the chronic conditions in the face of uncertainty. Chang and D’Zurilla (1996) report that a number of studies show that appropriate use of social problem solving is linked to psychological well-being, adjustment, and adaptive coping. Therefore, exploring and identifying the problem-solving style and skills of parents of children with LQTS will provide information on how well parents adapt to the chronic illness, as well as how they cope with the perceived vulnerability of their children and the uncertainty of the course of the disease. D’Zurilla and Nezu’s (2007) model of social problem solving will be considered to explore the effectiveness of parents’ coping abilities through social problem solving as they contend with their children’s LQTS and its unpredictability.

Components of social problem solving. D’Zurilla and Nezu (2007) describe two main components of social problem solving as it relates to coping associated with chronic and/or medical conditions: the *problem orienting response,* which is the metacognitive and motivational process of problem solving, and the *performance level,* or the individual’s actual problem-solving skills or ability, which is demonstrated by three basic problem-solving styles. *Rational problem-solving* is associated with positive problem orientation and an effective problem-solving skill set resulting in effective adaptation to the problem (D’Zurilla & Chang, 1995). The i*mpulsive/careless* style and *avoidant* styles are associated with a negative problem orientation and result in maladaptive coping, poor problem-solving skills, and ineffective behavior (D’Zurilla & Chang, 1995).

An individual’s orientation toward a problem is the motivational element, which encompasses the person’s operational set of cognitive-emotional schema that are oriented or skewed towards a negative or positive problem perspective. A positive problem orientation is defined as a set of cognitive operations or schema(s) that disposes a person to a constructive approach of problem-solving (D’Zurilla & Chang, 1995). This is demonstrated by the individual’s ability to (a) accept the fact that difficulties are an aspect of life and that it is possible to cope with situations in an effective manner, (b) acknowledge the problematic situation at hand, and (c) resist acting impulsively to the problem or in an avoidant manner (D’Zurilla & Goldfried, 1971). Those with a negative orientation are oriented towards an inhibitive cognitive emotional set that exhibits itself by (a) perceiving the problem as threatening, and thereby capable of undermining one’s psychological and social wellness, (b) demonstrating self-doubt in one’s ability to solve the problem (low problem-solving self-efficacy), and (c) displaying a sense of being overwhelmed or frustrated when presented with a problem (low frustration tolerance), leading to maladaptive coping behaviors (D'Zurilla and Nezu, 2007).

Empirical Support for Problem Orientation

Numerous studies have been conducted exploring the effectiveness of a positive problem orientation and coping. In 1986, Cormier, Otani, and Cormier demonstrated that individuals who were taught and trained to have a positive orientation to problem solving performed better on problem-solving tasks than those in a control group. Kant, D'Zurilla, and Maydeu-Olivares (1997) explored the relationship between problem orientation (along with social problem solving) and daily stress leading to depression and anxiety in middle-aged and elderly individuals of a residential community; they found that a negative problem orientation had a mediational effect. Those employing a negative problem orientation were more likely to be depressed and anxious, while those who employed effective problem solving along with a positive problem orientation demonstrated a reduction in emotional distress. In addition, a study on a nonclinical adult population by Chang and D'Zurilla (1996) demonstrated that positive problem orientation predicted active coping with daily stressors.

With regards to daily life stressors, researchers have found that individuals with a positive problem orientation exhibit more positive mood states (Elliott, Herrick, MacNair, & Harkins, 1994; Elliott, Sherwin, Harkins, & Marmarosh, 1995). In contrast, Elliott et al. (1994) found that a negative problem orientation led to more negative mood states in daily stressful situations, thereby suggesting negative trait affectivity. Individuals with a negative problem orientation are also shown to have increased worry and anxiety (Belzer, D’Zurilla, & Maydeu-Olivares, 2002; Bond, Lyle, Tappe, Seehafer, & D’Zurilla, 2002; Kant et al., 1997, Nezu et al., 1999).

Problem-Solving Styles/Skills

A rational problem-solving style involves a process of sequential skill sets, which studies support as an effective coping style (D’Zurilla & Nezu, 2007; Hill-Briggs, 2003). This skill set is best defined as a constructive problem-solving pattern that is rational and deliberate and exhibited by a systematic application of problem-solving skills (D’Zurilla & Chang, 1995). The skill set includes the individual’s ability to (a) define and formulate the problem, (b) generate a number of alternative solutions, (c) activate decision-making processes, and (d) implement a solution and verify its effectiveness, thereby making any changes that need to be made (D’Zurilla & Nezu, 2007). Studies have demonstrated that individuals who possess these skills are able to implement effective solutions to problems and cope more effectively with difficulties and crises, including those of a medical nature (Chang & D’Zurilla, 1996; Nezu et al., 1999; Noojin & Wallender, 1997).

The impulsivity/carelessness style is an ineffective problem-solving pattern. This style is characterized by an individual who attempts to solve problems actively, but tends to be limited in the generation of solutions, frequently employing the first idea that comes to mind. In addition, this person tends to display an impulsive, careless, and hurried (D’Zurilla & Nezu, 2007) attitude and behavior towards the problem.

The avoidant style is another ineffective problem-solving pattern. This pattern is demonstrated by an individual who procrastinates, becomes inactive and withdrawn in the face of difficulties, or exhibits a tendency to become dependent on someone else to deal and cope with the problem. Such individuals may also portray a denial of the problem, hoping that somehow the problem will solve itself (D'Zurilla & Nezu, 2007).

Empiricism and Problem-Solving Styles

A significant number of studies have demonstrated empirical support for problem-solving skills in decreasing depression, stressful life events, distress in relation to medical disorders, and other psychosocial problems. For example, Nezu, Nezu, Saraydarian, Kalmar, and Ronan (1986) discovered a relationship between negative life events and depression among college students. The more problem-solving ability the individual possessed, the less often depression occurred from negative life events. These findings were later replicated in individuals with major depression, demonstrating similar results (Nezu, Perri, Nezu, & Mahoney, 1987).

In 1995, Nezu et al. conducted a study on the effects of effective social problem solving in cancer patients. They found that cancer patients who exhibited poor social-problem-solving ability reported higher levels of depression and anxiety (Nezu et al., 1995) than those who exhibited more skilled problem solving. Among the medical population, individuals who exhibited better problem-solving skills also reported a higher sense of well-being. For example, Elliot, Shewchuk, Miller, and Richards (2001) discovered that individuals living with diabetes who are effective problem solvers report a greater sense of well-being. Grant, Elliot, Newmann-Giger, and Bartolucci (2001) conducted a study demonstrating the same correlation between effective problem solving and well-being among adult caregivers of individuals who had suffered a stroke.

Parents whose children have recently been identified as having LQTS suddenly find themselves not only in a parental role, but also in the role of medical caretaker for a child with a chronic illness. Social problem solving has demonstrated its effectiveness as an adaptive coping mechanism for others in the caretaking role of individuals with chronic disease. Caregivers of patients with spinal cord injuries demonstrate more effective coping when utilizing problem solving (Elliot & Shewchuk, 2003; Elliott, Shewchuk, & Richards, 1999). Rothenberg, Nezu, and Nezu (1995) demonstrated a similar correlation with caregivers of individuals suffering from Alzheimer’s disease. Finally, Kurylo, Elliott, DeVivo, and Dreer (2004) found that caregivers who display more effective problem-solving skills for family members suffering from congestive heart failure tend to be less depressed and exhibit less negative affectivity than those with less developed problem-solving skills.

CHAPTER 3

Method

The current study used a qualitative design and was conducted in a semistructured interview format. Grounded theory analysis, a method of qualitative research, was employed to explore the following phenomena: (a) how mothers cope with a diagnosis of LQTS, (b) how they define coping and what types of coping processes are utilized, (c) which resources they find most helpful in coping and adjusting to their children's chronic condition, (d) the effects of the disease on their quality of life and psychosocial adjustment, (e) how they contend with the uncertainty and unpredictability of the illness, and (f) whether a relationship exists between problem-solving style and problem-response orientation and mothers' psychological and emotional adjustment to LQTS.

Qualitative research was selected as the method, with the aim of the study to discover systematic concepts and relationships among patterns in raw data collected from the interviews (Creswell, 2007). Qualitative research is an inductive process which systemically generates theory from the data (Glaser, 1998). Strauss and Corbin (1998) describe qualitative research as a process that produces information about a person's life or other social-cultural phenomena not necessarily derived by statistical procedures. For the purpose of this study, the goal of utilizing qualitative research was to describe, interpret, and understand (Strauss and Corbin, 1998) the various biopsychosocial factors and patterns associated with the participants' subjective experience (Kadzin, 1998). Through this process, each participant's experience was recorded in detail, thereby providing new information regarding cognitive-behavioral and affective processes that otherwise may have gone undetected.

Following Strauss and Corbin’s (1998) model of qualitative research, three primary components to the qualitative process were followed. The first component was to collect data from the participants through detailed interviews to identify key cognitive, behavioral, and affective themes and patterns within and among the participants. The second procedure was to interpret, analyze, and organize the data derived from the interviews, which was accomplished by conceptualizing and reducing the data from the interviews into categories based on the properties and dimensions of subjects’ statements and descriptions. Finally, a coding procedure was implemented in order to enable the investigator and his team to derive systematic patterns from the data to compare, contrast, assimilate, and evaluate the meaningfulness of those patterns.

A more specific form of qualitative research is grounded theory. This research incorporated the fundamentals of grounded theory, which means the investigator did not begin with any specific or particular theories (Creswell, 2007). A general or “mean” theory evolves out of the data systematically gathered and analyzed through the procedures described earlier. As a result, grounded theory provided the opportunity for newly founded insight into and understanding of the subject matter while simultaneously allowing the investigator to set aside any preconceived bias and hypothesis (Strauss & Corbin, 1998).

Setting

The study data was primarily collected at the home of each participant. One participant was interviewed at her place of employment in a private office. The sample of participants was recruited from an electrophysiologist at a fully accredited and licensed hospital in the greater Philadelphia, Pennsylvania area. The interviews were scheduled at an agreed-upon time between the participants in the investigator.

Participants

Recruitment

A sample of 10 biological mothers who had children 19 years of age and under with LQTS was recruited from a local electrophysiologist (EP) specializing in treatment of LQTS patients. The EP selected 29 mothers of children with genetically confirmed LQTS from a list of patients. The list was provided to the investigator, who sent an informational flyer (Appendix A) with an introductory letter describing the details of the study to the potential participant (Appendix B). In addition, a stamped envelope addressed to the investigator and a response card were included. The participant was asked to check a yes or no on the response card indicating their interest in participating in the study. They were instructed to return the card in the stamped envelope. If they checked yes, they were informed that they would be contacted by the investigator to arrange a time and location convenient for them to complete the interview. If they responded no, they were told they would not be contacted. Finally, the introductory letter also stated that if there was no response or no card returned, the researcher would attempt to contact them by phone 14 days later. Of the 25 potential subjects contacted, 10 agreed and participated in the study.

Inclusion criteria. Participants were required to have a child or children 19 years of age or younger with confirmed LQTS. The mother(s) could be single, divorced, or in a heterosexual or homosexual marriage and/or partnership. The mother(s) needed to be capable of providing informed consent. The mother(s) identified themselves as the primary caretaker, which was defined as the individual who assumed primary responsibility for the child's care needs (activities of daily living), administration of medications or cardiological monitoring, emotional support, cardiac appointments, and homework or school activity participation.

Exclusion criteria. Mothers who were unable to read or understand the informed consent were excluded. Any mother who was unwilling to participate or who demonstrated undue duress about the interview process was eliminated. Finally, anyone who had suffered an LQTS-related death within the immediate family in the past 6 months was not asked to participate.

Characteristics

A demographic data form was completed by each participant after signing the informed consent and before beginning the interview (Appendix C). A total of 10 interviews were conducted. All 10 mothers were married, White, and had a combined household income of $75,000 or above annually.

The women ranged in age from 35 to 51 years old, with the mean age being 44.6 years old. Six of the mothers had bachelor’s degrees. Two of the mothers had high school educations. One had a master’s degree. One had an associate’s degree. All of the women lived in suburban or rural areas. Four of the mothers had two or more children with LQTS, and the remaining mothers had 1 child with LQTS. At the time of the study, the ages of the children with LQTS ranged from 3 to 19 years old, with the mean age being 11.2 years old. The age at which LQTS was diagnosed in the children ranged from birth to 16 years. Of the 14 children with LQTS, four had the condition diagnosed shortly after birth. Only three of the spouses had long QT syndrome. Tables 1 and 2 (below) provide more information on the mothers’ demographics and on LQTS in the children and other family members.

Table 1.

Summary of Participant Demographics

Pseudonyms	Age	Educational level	Mother's LQTS type
Lucy	43	Bachelor's	1
Betsy	51	Bachelor's	N/A*
Christy	37	Bachelor's	2
Nancy	45	Associate's	Unknown
Sally	46	High School	N/A
Gloria	46	High School	Unknown
Mattie	35	Bachelor's	2
Lily	51	Bachelor's	1
Tori	47	Master's	N/A
Mary	45	Bachelor's	Unknown

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* *Note.* N/A = Not applicable.

Table 2a.

Summary of Family LQTS Status and Symptoms

LQTS status of spouse	Spouse's LQTS type	Child(ren)'s LQTS type	Age at which LQTS was diagnosed in child(ren)	Age of child(ren) at time of study	Symptomatic child(ren)	Suspected symptom(s)	Age first symptom occurred	Number of incidents	Other family members with symptoms
Negative	N/A[*]	1	9 years	13 years	Likely	Syncope	7 years	2	Father, cardiac arrest
Positive	3	3	6 years, 8 years 11 years	14 years, 16 years 19 years	No	N/A	N/A	N/A	N/A
Negative	N/A	1	Birth	3 years	No	N/A	N/A	N/A	Mother, syncope or cardiac incident

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[*] *Note.* N/A = Not applicable.

Table 2b.

Summary of Family LQTS Status and Symptoms (cont.)

LQTS status of spouse	Spouse's LQTS type	Child(ren)'s LQTS type	Age at which LQTS was diagnosed in child(ren)	Age of child(ren) at time of study	Symptomatic child(ren)	Suspected symptom(s)	Age first symptom occurred	Number of incidents	Other family members with symptoms
Negative	N/A	1	3 years	9 years	Yes	Syncope	3 years	1	Mother, moderate symptoms
Negative	N/A	1	Birth	8 years	Yes	Arrhythmia	3 days	1	N/A
Negative	N/A	1	6 years	13 years	Yes	Arrhythmia	6 years	1	Mother, seizures and cardiac incident
Negative	N/A	2	Birth	5 years 7 years	No	N/A	N/A	N/A	Mother, arrhythmia
Negative	N/A	1	16 years	19 years	No	N/A	N/A	N/A	Mother, syncope

Table 2c.

Summary of Family LQTS Status and Symptoms (cont.)

LQTS status of spouse	Spouse's LQTS type	Child(ren)'s LQTS type	Age at which LQTS was diagnosed in child(ren)	Age of child(ren) at time of study	Symptomatic child(ren)	Suspected symptom(s)	Age first symptom occurred	Number of incidents	Other family members with symptoms
Positive	Unknown	2	Birth	4 years	No	Syncope	10 years	1	N/A
			10 years	18 years	Yes		N/A	N/A	
Positive	Unknown	2	4 years	5 years	Yes	Syncope	4 years	1	N/A
			14 years	15 years	No		N/A	N/A	

Protection of Human Subjects

Permission to conduct the study was obtained from the Institutional Review Boards (IRB) of the participating hospital and of the hosting institution. The investigator explained to the participants the goals of the study, the potential risks, the protection of the subjects' privacy, and the right to withdraw from the study. The informed consent described in detail the purpose of the study and the potential risks and benefits of participating (Appendix D).

Participants were afforded two initial occasions to decline to participate in the study and the opportunity to end their participation any time throughout the interview process. The first opportunity was during the initial solicitation by the investigator; the second was in person when the investigator reviewed the consent form with the participant. At that point and prior to initiation of the interview, the final written informed consent was obtained from the participant and witnessed by the interviewer. In addition, the investigator was also available and present to answer any questions or to clarify any confusion the participant may have had. Finally, a resource list regarding LQTS resources, counseling services, organizations, and support systems was provided to the participants at this time (Appendix E).

Participants were also orally told that this study was voluntary and that they did not need to participate in the study. In addition, it was orally explained and written in the consent form that the participants could terminate their participation at any time with no detriment to their medical care.

The informed consent included the participants’ consent to be audiotaped. The participants were advised of their confidentiality and that any identifying information would be omitted from the transcripts and protected from the public at large. In addition, the participants were informed that demographic data forms, tapes, written transcripts, and consent forms would be stored in separate locked files and destroyed at the appropriate and specified time.

Procedure for Data Collection

Mothers who were interested in participating in the study were contacted by the investigator. The purpose of the study was explained and the interview process was described. A convenient location for the participant was arranged. The investigator and participant met in person. After the informed consent was signed, a semistructured interview with the participant was completed. During the interview, a formulated list of questions was asked (Appendix F). Follow-up questions were also developed and utilized for clarifying participants’ answers.

Each interview lasted approximately 1 to 1½ hours. The audiotaped interviews were then transcribed by the investigator verbatim. Demographic information was obtained prior to the interview. Medical records were not reviewed for this study. Finally, each mother was given two free movie tickets for participating in the study.

Analysis of Data

Grounded theory was implemented in order to analyze the data that emerged from the interviews. A validation team was formed. This team consisted of one doctoral level psychologist, one doctoral level clinical psychology student, and one master's level psychologist who is also a doctoral student in clinical psychology. The team incorporated a triangulation strategy (Creswell, 2007), which corroborated evidence from the interviews, discussions, and coding, to confirm the validity of the study. Each team member was given the transcripts to review, along with a copy of the interview questions. Actual recordings of the interview were also available to the team members, if requested, to clarify any unclear information. Two transcripts were reviewed every 1 to 2 weeks. After every second transcript was coded individually by each team member, and a group discussion or dyad was facilitated in order to collaborate, confirm, and validate findings of the transcripts read. At each meeting, the verification team reviewed and ensured that content was accurate and confirmed the evolving generation of the hypothesis and patterns in order to shed further light on any perspective themes (Creswell, 2007). After all 10 transcripts were analyzed, the three coders involved gathered together to do a final analysis of contrasting, comparing, confirming, assimilating, and processing the themes to validate the final results.

Each member spent 2 or more hours examining and coding each transcript independently. Additional time was spent on a weekly basis working collaboratively as a team. During each meeting, the investigator took scrupulous notes of each team member's thoughts, analyses, reactions, and coding processes.

CHAPTER 4

Findings

Diagnosis: The Dawning of Consciousness

Of the 10 mothers interviewed, four had prior knowledge of LQTS before it was diagnosed in their child, due to their own LQTS. These mothers discovered LQTS as a result of their own LQTS incidents and symptoms. Although these mothers had some concept of LQTS, identification of their children having the syndrome generated an elevated consciousness. It appears that in some manner, LQTS had a greater impact when it was diagnosed in their children. Five other mothers had never heard of LQTS nor had any conceptualization of it until their children exhibited symptoms and it was diagnosed. Finally, the 10th mother became aware of long QT as the result of her husband's cardiac arrest. None of the mothers interviewed had children who had died from LQTS.

Although the majority of mothers reported being satisfied with their children's current medical care, 50% of the mothers had a difficult experience with the medical community upon learning their children had LQTS. For example, as will be discussed later, these mothers reported their initial interaction with health care workers as confusing and ambiguous because LQTS was not explained fully or they felt it was treated too lightly. A few reported that some health care workers did not fully appear to comprehend LQTS. Others recounted that the staff appeared less than sympathetic. One mother's LQTS was misdiagnosed. In short, it appears these experiences added to the mothers' distress.

The other 50% of mothers reported either a positive experience with the medical staff upon learning of LQTS or made no mention of it. It is noted, however, that the majority of the mothers who had an initially difficult time eventually found professional medical services which either evolved into more positive relationships with the health care staff or changed medical facilities.

Mothers Who Knew They Had LQTS Prior to the Diagnosis in Their Children

Please note that some characteristics of the participants and their children were changed to protect their identities. For example, gender was reversed for some of the children. Some of the actual descriptions of locations and activities were altered to protect identities. Finally, all of the mothers and children were given pseudonyms.

Christy's story. Christy's LQTS was diagnosed when she was in her first trimester of pregnancy with her first son. Christy stated she discovered that she had LQTS due to either a cardiac episode or syncope while at home. She wasn't quite clear during the interview as to what actually occurred. She stated that she had severe hyperemesis (excessive vomiting) due to her pregnancy. On one particular day, she stated, she was feeling very ill. She had called her obstetrician for an appointment. She stated, "We drove all the way to Pennsylvania. On the way home, it was a snowy day. Anyway, a long story short, I passed out. I was taken to a local hospital in my state. I found out that I had something called long QT syndrome."

Christy did recount that a few weeks earlier, her cousin called her to tell her that she had long QT syndrome. "And honestly, it went in one ear and out the other. I knew there was something serious about it and she was on something called beta-blockers. But honestly, I did not remember the name of the condition. So, the puzzle started to be put together. Here I am in a hospital. I call her up and I said, like ‘Susan, what was the condition again?’ She said, ‘long QT syndrome.’ My doctor came in. I said ‘look, like, send off for genetic testing.’” Another complication was that Christy was pregnant. She stated that due to being pregnant, nobody really knew what to do with her. “So I had to stay in the hospital for weeks and until I was out of my first trimester. As soon as I was out of my first trimester and I was 15 weeks pregnant, they implanted a defibrillator. . . . As I looked into this disease, I learned there was a 50% chance of passing this genetic mutation onto the baby. When the baby was a week old, they diagnosed him with LQTS. Without saying, I was devastated. He was only a week old. I guess there was a very strong level of denial knowing intellectually that I had this genetic mutation.”

Nancy’s story. Nancy, a mother of two, learned she had long QT syndrome a year prior to the diagnosis in her son’s LQTS. Nancy discovered her diagnosis of LQTS through a cardiac episode which she describes as a sense of “impending doom” and “death.” At that time, “the doc said that I was borderline QT.” Nancy was put on beta-blockers and carried on with her life without much regard for LQTS. Approximately 6 years ago, when her son was 3½ years old, he had his first episode. Nancy was hosting a gathering at her home. Her neighbors were there, and the children were playing. Shortly before her son’s episode, she stated that he was swimming. He was then resting and watching the other children play. Her older daughter, who was 15 years old at the time, was watching the younger children. “I was on the porch so I was not real far away. I was kind of peeking out of the corner of my eye. He just said, ‘My belly.’ And she (my daughter) said, ‘Come here.’ He started to walk towards her. He just fell flat to the ground. Of course, like I saw part of it. She yelled out. I ran over. She basically handed him to me. He was totally limp. He was unresponsive. He was pale and his eyes were wide open. He was out. He was out. He was like a little rag doll. I was screaming." Nancy's son was taken to a pediatric hospital by ambulance. The mother did not disclose whether her son had a cardiac arrest or had an LQTS syncope incident. However, she did disclose that her son was later found to have long QT 1 and long QT 3.

Mattie's story. With regard to the interviews, Mattie appeared to report the most benign reaction regarding her children's LQTS among the mothers. Mattie, who was in her early 30s, first learned she had LQTS at the age of 14 years. "You know, having to face your mortality at age 14 when you're invincible was an extremely hard concept." A few years later, when she was 17, Mattie experienced "sustained ventricular tachycardia for 72 hours. . . . I was on the Holter monitor. They rushed me into the ER. The electrophysiology lab. They decided the best action for me was an implanted defibrillator."

Mattie described that she had adjusted and coped with long QT syndrome throughout her late adolescence and young adult life. Therefore, she knew before she decided to have children that they would be at risk for LQTS. Both of her children have type 2 LQTS. Mattie stated that accepting and coping with her children's LQTS was not particularly hard. She stated that because she has dealt with this for such a significant length of time, it no longer felt abnormal to her. However, what she did find disconcerting was the concept of possible sudden death of her children which she admitted was "worrisome" and "difficult to overcome."

Lily’s story. Approximately 3 years ago, Lily, who was 49 years old at the time, learned she had LQTS 1 due to several incidents of passing out while she was involved in strenuous physical activities. “You are feeling fine. Then suddenly somebody tells you that you can’t do anything. You know, you can’t do this. You can’t do that. I passed out. I had an episode where I passed out. I was hiking high in the mountains. So I always thought it was the altitude. And then when they diagnosed me and asked have you ever passed out, yeah, well it was the altitude. They said, ‘No it wasn’t.’ That is why I have the defibrillator because I passed out.” As the result of Lily’s diagnosis, the rest of the family was tested and screened. Lily’s son was 16 or 17 years old at the time and was found to have LQTS 1. Results for her husband and other child were negative for LQTS.

Lily was direct about her noncompliance and her frustration over the illness. However, when it came to talking about her 19-year-old son, she shared feelings of guilt and deep concern. Lily’s primary concern with her son was that her son was also noncompliant with treatment protocol, especially since living away from home. This resulted in Lily’s inability to have more control to monitor her son’s daily routines.

Mothers With No Prior Knowledge of LQTS With Symptomatic Children

Lucy’s story. One mother, who will be identified as Lucy, described having a son who had a series of serious falls. The first fall occurred when the son was quite young. He had fallen off a high ledge. As a result, he broke numerous bones and needed surgery. At the age of 10, he had another serious fall likely due to loss of consciousness. He was then taken to a local hospital.

Lucy elaborated on her own hospital experience by explaining her frustration with the emergency room physician. She stated he negated the full ramifications of her son’s fall. “We take him to the hospital. They x-ray his head and all the stuff they would do when you become unconscious. And the doctor in the ER said ‘it is a moot point as to whether he passed out and fell or fell and passed out.’ I said, ‘I don’t think that is a moot point.’” Lucy, being distraught over the physician’s reaction, decided to contact her pediatrician. Lucy reported that her pediatrician stated that it wasn’t anything to worry about and that she “was not entertained by the incident.” In fact, the pediatrician suggested that if she was concerned, she should contact a neurologist because the son probably had a seizure. However, given Lucy’s persistence and concern, she decided to talk to a friend in the medical field. Her friend suggested that she should have her son examined by a cardiologist. Due to Lucy’s concern and determination, they discovered that Lucy and her son have type 1 long QT syndrome. However, her husband and daughter do not carry the gene. She credits her friend for directing her to the appropriate resources.

Sally’s story. Sally, a mother who generally had a positive experience with the medical community, stated, “When my son was born, the pediatrician told me he thought he heard an arrhythmia in his [son’s] heart. He was 2 days old.” An electrocardiogram was performed. The pediatrician sent the EKG to a pediatric hospital for a consult. Upon a medical screening of family history and additional testing, the diagnosis was long QT syndrome. Sally reported taking the news of the diagnosis in stride initially. She also talked about her “love for” and “trust in” the cardiologist, who she believed assisted her with understanding and explaining the syndrome. She also suggested that because her first son, who is 2 years older, was born prematurely and had other complications, she was prepared in advance. In other words, Sally felt that the trauma of her first son assisted her in preparing for the challenges of her child who has long QT.

Finally, she mentioned that because she didn't understand the full ramifications of LQTS at the time, and because her son was so young and more easily monitored at first, the diagnosis was less disturbing. It wouldn't be until a short time later that her distress about the disease increased. For example, as the seriousness and randomness of the disease emerged within her consciousness, she was no longer to able to discount the disease as something her child may grow out of one day.

Tori's story. Tori, a mother who has a son and daughter, both with LQTS, found out about her son's long QT syndrome through an incident at school. At the time, the son was 10 years old. While he was playing in the school band, he felt as though he was going to pass out. He left the activities and started walking to the nurse's office and fainted. He was sent to a local hospital, where LQTS was diagnosed. Her son was in 12th grade at the time of the study. "Once we consulted the doctor, we became aware that it [LQTS] was called sudden death or arrhythmia." Four years ago, the mother gave birth to a daughter, who also has long QT syndrome. As with Sally, Tori's medical experience appeared to be more informative and she appeared to have a trust with the professionals she was dealing with and continues to deal with.

Mary's story. Mary described a difficult enlightenment regarding LQTS. In the summer of 2007, Mary and her two sons were visiting family and friends. In the morning, the boys took their family dog for its morning walk. She stated that they went a short distance. "They just went out the side door, down the driveway, and across the street."

The next thing she stated was that, “my eldest son was carrying in my little guy. He was completely unconscious. He just carried him in. He was just completely limp. So, I just kind of freaked. My husband was out of town at the time. But my daughter was there and she kind of ran into the room. We just laid him down in the kitchen. I think my first reaction was that I freaked and I was screaming. He didn’t come to for about 10 minutes. He was in the ambulance when he came to.”

Initially, physicians at the local hospital were concerned about head trauma. The son was admitted for 24 hours. Mary continued her story by stating, “The next day when we were literally signing papers for discharge and a doctor walked in and then said, ‘We think there is a heart thing going on.’” Mary expressed frustration that the medical staff did not pick up on the arrhythmia from the EKG. She stated that it was her pediatrician’s office that noticed the long QT wave and faxed the EKG to a local cardiologist and the hospital.

Mary also described a sense of unconcern from the hospital staff and the cardiologist. “They sent us home. I think they may have sent us home with a prescription for beta-blockers and a little card to call their office. This happened on Friday. So, it was the weekend when we were being discharged. So they were like, call our office on Monday and in the meantime take the beta-blocker. So that was that.” Mary expressed frustration with regard to learning how she was informed about LQTS. As with Lucy, she implied and described an initially difficult time with the professionals diagnosing the condition.

Gloria’s story. LQTS was diagnosed in Gloria’s son while he was being treated for another illness. He was admitted to the emergency room at a local urban hospital for a high fever, fatigue, and joint pain. Medical staff decided to do an EKG to determine if he had rheumatic fever. It was at that time they discovered that her son had long QT syndrome. Gloria's son was 6 years old at the time. He was 13 years old at the time of the study and was being treated with beta-blockers.

Upon learning her son had LQTS, Gloria and her husband were screened for it. Gloria stated that the cardiologist said there was some concern regarding her status. However, she reported that the cardiologist concluded, at the time, that she was fine, but that she should eventually have it checked out. Her husband results were negative for LQTS. Several years later, Gloria and her family decided on genetic testing. Gloria's results were positive for LQTS.

Although Gloria's LQTS had been previously diagnosed, her first noticeable incident did not occur until 2 years ago, when she was approximately 44 years old. However, it would not be until a year later and seven visits to the hospital emergency room that her symptoms were finally linked to LQTS. She stated, "I woke up Memorial Day morning to my husband's alarm clock. The number 2 gene is noise. The alarm clock woke me up. I felt like I was dying. And I said that to the doctor. The doctor said that I was crazy. Literally I could not open my eyes. I could not do anything. And then it happened again at the hospital. They said that I needed to see a psychiatrist because they thought I was crazy and stressed out. But what happened was the long QT. I was diagnosed from a major hospital in South Jersey. I won't give you the name. They diagnosed me with syncope. Syncope is the first sign of long QT. Then I started having seizures. Then they put me on migraine medicine" (which she stated later is contraindicated for LQTS). Although Gloria was told she had syncope and a seizure disorder at the time, the link to LQTS was not made until she was referred to an electrophysiologist a year later, after suffering through several misdiagnoses.

Gloria’s own experience with LQTS left her with diminished confidence in the medical community. As the result of her unfortunate incidents and experiences with several misdiagnoses, she came to the conclusion that a significant number of medical professionals lack adequate knowledge of long QT and its symptoms.

Diagnosis of LQTS in Another Family Member

Betsy’s story. The dawning of Betsy’s knowledge of LQTS occurred in 2000, when her husband, who was 38 years old at the time, was leaving the house to walk to the local convenience store. He only got a short distance when he went into cardiac arrest. Emergency services were called. While CPR was being conducted, the police arrived with an external defibrillator. However, Betsy stated that they did not know how to use it. As a result, the police were later trained in the proper usage. Fortunately, her husband recovered “but within a week, they determine that he was potentially LQTS. Interestingly enough, he had a full physical with an EKG two weeks prior to this happening and the EKG was normal, or at least normal enough not to raise any red flags.”

After LQTS was diagnosed in her husband, it was recommended that the children be tested. At the time, the boys were 11, 8, and 6. All of the boys were genetically tested. The two younger boys tested positive for LQTS 3. Her older son was not positive according to the genetic test, but due to clinical symptoms he was being treated “prophylactically.”

Betsy appeared to be one of the mothers more frustrated with the medical profession. She expressed frustration regarding the ambiguity of the diagnosis. For example, she was not sure why her older son was being treated for LQTS. She believed there is a divergence of opinion among physicians and she suspects that scientific and medical information is being withheld from the patients. Betsy said her current physician has her sons on sodium blockers. She believed this may be too aggressive and believed her previous cardiologist would not have suggested the medication. Ultimately she felt she was receiving “filtered” information which is “whispered down the lane” from top researchers to the physicians. She was uncomfortable with the idea that the treating physicians do not have a consensus on how to treat individuals with LQTS. She believed she “should have access to all the best information,” which she suspected is withheld from the layperson.

Initial Reactions: 9/11 Shock and Awe, the Incomprehensible

“For the kids it was like, okay, here’s this medicine you can take. But at the same time, it could happen, whenever, to your kids. So at first, it kind of reminds me of not long ago, after 9/11 happened. Right after 9/11 you feel very vulnerable. Like any given moment, any given day, thousands of people could be killed . . .”

The following themes emerged from the mothers’ discussion of their reactions to hearing and contending with the diagnosis of LQTS. A word count of adjectives describing emotional states was conducted to determine the frequency of word usage through each of the interview transcriptions. The word count, along with a tally of how many mothers endorsed each of the various emotional states, is presented in Table 3. For example, all 10 of the mothers used a combination of terms such as *fear, scared, upset,* and *anxious* a total of 78 times throughout the interviews. *Denial* and *disbelief* were spoken 22 times by eight of the mothers. *Guilt* was addressed 14 times by three mothers. Eight of the mothers used the words *shock, devastation,* and *trauma* nine times to describe their experience of with LQTS. Finally, it should also be noted that the author and the validation team interpreted various phrases, sentences, and passages which identified emotional content and themes.

Denial/Disbelief

Eight of the 10 mothers, or 80%, described experiencing some level of either disbelief or denial regarding the diagnosis of LQTS in their children. As previously noted, the words *denial* or *disbelief* were used a total of 22 times throughout the eight interviews. For example, Lucy stated, “I didn’t believe it. There’s still a part of me that doesn’t believe it. I really don’t believe it. It seems so obscure. It’s nothing we’ve ever heard of and he’s a big hearty kid. It just doesn’t make any sense.” Christy relayed, “that is another thing about this condition. It really enables a high level of denial because you’re not looking at a child with Down syndrome or not looking at a child who’s got cancer. You look at these perfectly normal children who function very normally. Yet they are walking around with these life-threatening conditions. And for those who want to stay in denial, it really enables that in my situation.” Nancy stated, “I didn’t want to believe it at first. . . maybe he really doesn’t have this diagnosis. So, there was a little bit of denial

Table 3.

Rank order of word count/emotional endorsement by word usage

Emotion endorsed	Total word usage	Number of mothers who used descriptor
Fear, scared, upset, anxious	78	10
Denial/Disbelief	22	8
Guilt	14	3
Shock, devastation, trauma	9	8
Sad/Loss	10	4
Vulnerable	2	2
Anger/Frustration	2	2

that I went through." Betsy shared that she did not initially "believe the lab results." Lily was also forthcoming in her admission of denial, "Yeah, I wish I would never have found out about it. I don't believe it; I don't buy that it is as deadly as they say it is. I guess I'm in denial."

However, Sally's denial was not so quick to emerge to consciousness. She reflected that she was attributing LQTS to something doing with the birth of her son, "I don't know if you know, but there is some theory that he was so young that it takes 6 weeks before the heart system really takes off on their own. Okay? And hope was always there and that Timmy being premature and his age had something to do with that. That cardiac problem. Not, not that he truly had it. So in my head, I think that is what I heard as the mother. You know what? He is going to be okay. I really thought God was going to watch out for him."

Although Betsy witnessed her husband going into cardiac arrest prior to the diagnosis in her children, she discounted the seriousness of the illness by stating, "I probably have more of a chance of driving up to the grocery store than having something happen to these kids with this" (referring to LQTS). Gloria unintentionally neglected to take the diagnosis seriously. It was not until a few years later and experiencing her own cardiac incidents that she broke through denial about the illness. Mary simply stated that she did not remember her initial reactions. It may be possible that Mary mentally blocked out portions of her initial experiences with her son's LQTS incident.

Regarding the two mothers who did not use the term denial, it does not mean to imply that they did not go through a moment or time of denial. Mattie has been coping with her own LQTS from a young age and knew the risk of passing LQTS unto her children. She also had relatives with long QT syndrome. LQTS was not a new concept for her when it was diagnosed in her children.

Discovering and experiencing her son's LQTS was a new experience for Tori. However, she tended to minimize her emotions throughout the interview. When she did express a difficult emotion, Tori tended to retract it. It is possible that she may have wanted to present herself in a more favorable light for the interviewer or that she was still keeping her underlying feelings at a distance.

Shock, Devastation, and Trauma

In addition to a sense of denial, numbness, or disbelief, 6 of the 10 mothers endorsed feelings of shock, devastation, and trauma. “This was shocking. We were really freaked out. But it catches your attention that you find out that people die of this and that usually the first incident that they know about is death” (Lucy). Christy recounted, “I was devastated. It felt like a death sentence. That phrase, sudden cardiac death, is a heavy term to embrace. I felt in many ways traumatized. . .” Nancy went on to say, “This event was really dramatic to me.” Even though Sally initially stated that she took the diagnosis in stride, she later went on to state in the interview, “the thought of losing your. . . that sudden death. Devastating. And you couldn’t do a damn thing about it.” Mattie, who learned she had LQTS as a 14-year-old, stated that it was shattering. And finally, an interesting response from Tori was that she initially stated, “Well, it was devastating.” However, later she went on to recant her statement and say, “Well, it was not that devastating.”

All of the mothers, upon hearing of the initial diagnosis in their child, expressed some kind of shock, awe, disbelief, denial, or other kind of traumatic experience. However, Mattie discussed her trauma or devastation about the diagnosis in relation to herself at the age of 14. She tended to describe her experience with her children as having lost the lottery. It is likely that she has had years to cope and adjust to the condition, whereas Tori initially described an intense emotional experience, but then backed away and discounted the initial reporting of her emotions.

Evolving Emotional States

It appears that all of the mothers had some initial distress upon hearing the diagnosis. Sally stated she took it in stride, but later admitted to developing fear and distress once the concept hit her. A majority of the mothers appeared to have initially reacted by incorporating some level of denial or numbing. Other mothers described their initial reaction as shock or devastation. However, a theme that did emerge throughout the transcripts and interviews was that all of the mothers eventually developed a state of fear and distress. This is likely due to the fact that as the shock and initial reaction wears off, the knowledge of the uncertainty and the randomness of sudden death begins to be assimilated. This stage ebbs and flows for many of the mothers. Some mothers appeared to have worked through much of their anxiety, whereas others tended to perseverate on their fear. However, as one mother stated, as time continues, one begins to feel more secure. Eventually, the majority of the mothers seemed to have learned to cope with the anxiety and randomness of the disease. However, given the right circumstances or situation, it was easy for the mother(s) to drift back into a sense of dread and fear.

Scared, Fearful, Distressed

All 10 mothers, or 100%, used expressions such as *fearful, scared,* or *upset* as their knowledge and understanding of LQTS evolved. These words were used a total of 78 times throughout the 10 interviews. For example, Mary stated, “It was really upsetting. And of course, as we learned more about it, it just got really scary; just really scary. I am afraid.” Lucy reported a sense of “fear hanging over [her] head.” Christy talked about having an ongoing fear and anxiety. “Living in the fear that something could happen at any given moment to my child, even on medication, scares the hell out of me.” Nancy summed it up by reflecting, “You know, like fear that you can lose them at any time. You know, at any given moment. You know, fear that the medication they are on is not 100%. That is the hardest part for me. I mean, there is a lot. I could go on.” Gloria stated, “You know, they can drop like that.” Mattie, in spite of her more benign attitude, stated, “Well, the only thing that is difficult is the possibility that they may go into cardiac arrest.... it’s uncontrollable.” Tori expressed her fear by contemplating the concept that “we became aware that it was called sudden death or arrhythmia. It is very random.” Sally went on to state simply, “it can be sudden death to him.” Betsy reported, “it is disconcerting, as a parent, knowing that kind of LQTS can happen at rest.” Christy continued, “It is like a gamble. That is exactly what it feels like.” Finally, Lily succinctly revealed, “it’s just that your kid gets it and it’s scary.”

Sadness/Loss

Although fear appeared to be the predominant emotion, four of the mothers, or 40%, discussed feelings of loss and or sadness. None of these mothers experienced deaths as a result of the LQTS. The loss and sadness seemed to be related to the restrictions that they perceived their child may have during his or her lifetime. For example, Christy clearly stated “I had a lot of sadness and a lot of fear and a lot of anxiety.” Christy reported that her sadness and loss was about her child’s restrictions from sports. Christy talked about how she was an athlete and how she participated in so many activities. To her, this aspect of participating in athletics brought such joy to her life that she talked about feeling guilty and sad that her son will never be able to experience the quality of life she has experienced. Lucy also described her feelings about the loss regarding her son’s inability to experience a life of athleticism. “And I was really saddened by the complication of exercise. This will affect him for the rest of his life. No stimuli; loss of the sports.” Mary lamented that her son will not be able to participate in sports. “You know, I just wished, I just feel sad. I just wish that I could bring him to soccer practice like I do my older son.”

Nancy, on the other hand, described her sadness simply by realizing that her son has long QT syndrome “in that first night lying next to him and, you know, I was crying,” Later she stated that a year ago, her son was watching football on television. She recounted, “And it was out of the blue. He just had tears in his eyes.” She asked her son, “like, what’s the matter with you? And he just said, ‘I just hate my heart problem because I will never be able to play football.’ But out of the blue he said that. And I was like wow. I said, ‘well, honey, you know there are lots of kids who can’t do at a lot of different things. You can do this, you can do that,’ you know, like I tried to show him how lucky he is in other areas. Because I have done that for myself. Everything that I have gone through, I am trying to teach him in terms of coping with the diagnosis.”

Guilt

For several of the mothers, guilt was a factor in their response to the diagnosis of LQTS. Three of the mothers actually used the term to describe their reaction. A few of the other mothers appeared to imply a sense of guilt. Lily was very direct about her feelings of guilt. “Oh, I felt horrible. I felt guilty. You feel guilty. It comes from my side.

Yep, I felt guilty. I felt terrible. Well, I think that any time you pass some sort of illness on to your child, you feel guilty. I felt terrible that he got it from me."

Another mother did not use the word *guilt* in her interview. However, she referred often to how she got away with things in her life. She talked about having played sports and relates about experimenting with an alternative lifestyle, which included recreational drug use, in her youth. She didn't quite comprehend how she was able to get away with it, since she has LQTS. However, she was afraid that her son will not. Christy was also direct about her feelings of guilt. She stated, "Yes, so I am the culprit here." Christy was referring to the fact that the LQTS gene was passed down from her side of the family.

Mattie appeared to express some form of guilt. Mattie talked about how she took the chance to have children and now they both have LQTS. Although she did not directly express guilt about this, she went on to talk about her aunt dying of LQTS. During that part of the interview, she addressed feelings of guilt that her aunt's sister still lives with.

The other mothers alluded to the possibility of guilt feelings. For example, several of the mothers talked about the loss of their children's quality of life by not having the same opportunity to participate in activities that they were able to do. They seemed to imply a sense of guilt that they had pleasures that their children will never have. However, because there does not seem to be direct evidence about this theme, they will not be included in the discussion about guilt at this time. But as one may read between the lines, especially those mothers who talked about the fear of their child not being accepted as normal, one could possibly perceive some element of guilt and/or shame about not producing a healthy child or lacking the control to protect her child from the disease. For example, Christy's demonstrated this point in her interview when she stated, “I had this beautiful newborn baby boy. I felt like, you know, I don’t know how to say this but it was like he was already damaged goods.” She continued, stating “so I just had this instinct to want to protect him and the fact that I cannot control this 100% is my biggest battle. This is whipping my butt. Like I can’t fix this; I cannot control this.”

Behavioral Effects and Perpetual Anxiety

As seen from the pattern of emotions that emerged from the mothers living and coping with LQTS, a significant behavioral pattern that emerged among many of the mothers was a hypervigilance and/or a preoccupation (or continual mindfulness) of their children’s safety and well-being. Nine of the 10 mothers expressed hypervigilance and a preoccupation with their children, especially when first learning of the diagnosis. One mother appeared not to have engaged in overmonitoring behavior or the preoccupation as much as the other mothers. It was this mother in particular who appeared to remain in a state of denial throughout the interview regarding her child’s LQTS. She consistently stated that she did not believe that LQTS was as bad as most people think. In addition, she believed that her child was “hardy” and not as much at risk for cardiac incidents as her physician suggested.

The other nine mothers referred to a perpetual anxiety, especially after first hearing of their children’s diagnosis. “You’re wondering, okay, how do you let them out of your sight or when they are not with you, especially as a mother, it’s like, how do I protect them and how to make sure that they are okay?” (Betsy). Christy mentioned, “it is always on our minds whether we are in a good frame of mind or not. Nancy stated, “and of course, I wanted to keep him in a bubble, you know, and stay with him all the time.” For Sally, it was a bit different. She related, “It was easy at first because he was an infant. He was always with you. He was with me 24 hours a day. You can control your environment. You can keep an eye on him.” However, Sally recounted a different story about the present, now that her son is older, “we watch him closely” Gloria reinforced this concept by stating, “so, there is a fear that you always have. I have to watch him. I always make sure that he is okay. I guess my husband and I both have gone into his room to make sure he is still breathing.” Mattie reinforced this by stating, “we still get up in the middle of the night and go in their rooms and check on them and make sure they are still breathing.” Tori confirmed this: “The more I got into research, you know, these kids would die in their sleep. When I read that, if he slept too long, I would go in and check on him and stuff like that.” Mary stated, “of course, in the beginning, it is almost like you don’t want to take your eye off of your kid. You’re looking for symptoms. And you don’t know if the medication is working. So in the beginning it was a lot more difficult than it is now.” Finally, Lily stated, “I worry. You know, I worry about if something bad happens. I worry about him.”

Behavior and Fear Management: How Mothers Have Changed Their Environments to Protect Their Children

Every one of the mothers engaged in behavioral engineering of their immediate environment. This included monitoring the children more closely, medical compliance, restricting their diets, curtailing and redirecting physical activities, and reducing other triggers within the environment that could potentially precipitate a cardiac episode. Also, the mothers’ urgency to modify the environment appeared to give the mothers a sense of control over what otherwise is an unpredictable and random disease. For example, Lucy takes “comfort in the fact” that she was doing everything that she believed she was supposed to do. “We just did what we were told to do. We just did it.” Betsy shared, “we are conscious of the alarms going off in the house. Controlling what I could control to the best of my ability. The list is just ongoing.” However, Betsy was also cognizant that she cannot control everything. “Can you choose your alarm to not be a jarring alarm? Yes. But you tell them, you know, it’s probably best if you don’t try to startle each other and do things like that. Can you control every little thing that happens? No.”

Christy talked about how she had programmed herself to stop her children from running about the house and being too rambunctious and to redirect her children’s physical activities. Nancy reported becoming aware of various triggers in the environment. She talked about attempting to monitor her son’s emotional stress. Sally was vigilant about informing other people about her son in case an incident would occur. Gloria made sure her son did not overdo it physically when she was around him. She also monitored his sleep habits and diet. Mattie talked about becoming more creative with physical activities and planning play dates that did not involve sports. Tori addressed the routine of medicine and monitoring her older son by being sure that he always had a cell phone with him.

Two of the mothers did report learning CPR. In fact, one of the women teaches CPR in the schools and community. Several of the mothers also had AEDs. Each mother appeared to have their own comfort level as to when, where, or which activities to take the AEDs. Some of women who have learned CPR and who had AEDs expressed having stronger peace of mind as a result.

Lily, on the other hand, expressed more concern and feelings of being out of control. This was because her 19-year-old son was living at college and she realized that he was noncompliant with the medical recommendations. She stated that all she could do was nag him. Finally, Mary expressed feeling badly because she made her younger son sit in front of TV most of the day. She also expressed concerns about her older son because she didn’t have the ability to monitor him as closely.

Social Systems and Institutions

Social Milieu

Coming out: To tell or not to tell. As mentioned in the previous section, all the mothers had concerns about making the environment safer for their children. However, regarding the social system, several of the mothers had difficulty sharing about their children’s chronic illness with friends and neighbors. The mothers appeared to be concerned about social stigma and others' perceptions of their children's vulnerability. A few of the mothers expressed a fear of their children being ostracized from the social community. Several mothers expressed concern that their children would be treated differently or unfairly and not be considered normal like their peers.

For the mothers who had children who were in their adolescence, two of the adolescents in particular wanted to keep their LQTS secret. The other concern that mothers contended with was the inability to control others' emotional reactions. For example, Lucy stated, "people get scared. What you don't want to do is you don't want to scare people. I remember my best friend in eighth grade was epileptic. My mother didn't even want her to come over. You certainly can't drive with them and that sort of stuff." Betsy stated, "I didn't want to have to tell a lot of people . . . I mean, it was hard because I didn't want to worry those people. I didn't want to say, when my kids are over to play, something terrible may happen and at the same time I didn't want them to know that something had changed." Betsy also struggled as to when to take the AED with them on trips and vacations. She seemed to only want to take it when she thought they would be far from any hospital or medical facility.

Mary expressed fear about her child being ostracized. Christy expressed concern as to how she was to represent this to the public. She was afraid of undermining her son's well-being by frightening other people about his vulnerabilities. "I have seen people look really nervous after I talked to them and they are a little afraid to be with him alone," whereas Nancy stated, "so, it gets like that social aspect of how to work this into your life. Who do you tell? So I tell every friend that he goes to play with and I explain to the parents that he has LQTS. You have to tell everybody." For Sally, she reported "I think the hardest thing to accept at first you have to tell people, I feel."

With Gloria, it was not so much her concern about telling others as much as it was her son's concern. Her son, in whom LQTS was diagnosed when he was 6 years old, was 13 years old at the time of the study and didn't want anybody to know that he was different or that he had a chronic illness. It was not discussed whether her son was less concerned about others knowing when he was younger. Gloria felt that she had to disguise her son's LQTS and could only tell a select few of his friends and their parents in order to avoid her son's anger towards her. Mary found herself in a similar place as Gloria. It was difficult for her to be public about her adolescent son's LQTS, which was diagnosed when he was 14. She stated that her son became very angry when she informed others. Therefore, she had to be covert when telling others in order to avoid upsetting him.

For Mattie, she believed one of her biggest challenges was the social aspect. She felt it was easier for her daughter because she's female. For her son, she felt it would be more difficult because of the sports involved. "I've had to explain to the other mothers again, you know, part of that social aspect. But not all the mothers know. If they ask, I will quickly say something. So basically if it is a necessity, if people ask, I let them know what is going on."

Lily didn't speak much about the social aspect of LQTS. She did mention that her son tended to use the condition to his advantage. She explained that her son had used it to get out of hazing in college and other situations. In other words, he used it as a "sympathy card."

Tori claimed that she and her family did not hold back from letting people know about LQTS. They didn't seem to be as concerned about the social stigma. It appeared that her concern was that other people do not understand LQTS. "They [referring to friends and parents] just assumed that if he took the medication that it would cure him, especially the sports parents. They would think that he would not be limited and things like that. They would see him out there. He looks normal. He looks like he has no restrictions or anything at all. We explained to them. They were fine. It has been such a long time that his close friends and their parents all know about it and are very accepting of it."

In addition, several mothers shared concerns about friends and acquaintances not grasping the ambiguity and full consequences of LQTS. They feared there is a lack of public awareness and misconceptions about the disease. This concern frustrated these mothers and was accompanied by a sense of being isolated from the community at large. Betsy exclaimed, "Yes, I am finding that out with people. And you will find that out. You will see. They don't understand the seriousness of it. Another mother (Gloria) went on to state, "you have to discriminate with whom he stays with. Some of the people weren't responsible as parents." Mary stated, "People don't take this all that seriously as to what it is."

School System

Two of the mothers had difficulty negotiating the school system. Lucy believed that the school system overreacted. She reported that the school wanted to take precautions that she did not perceive as necessary. She believed the school was too reactionary. She mentioned that she believed the school administration was primarily concerned about legal action not about her son. Lucy also mentioned that the school wanted her son to carry an AED with him. Lucy stated, “Fat chance. The first day of school was horrid.”

Mary also had difficulty with the school system. She expressed frustration about the school’s demands regarding field trips. She stated initially they would allow her to accompany her son. Later in the year, they did not permit her to go. They insisted that the son have a nurse escort him. Mary felt this was unnecessary. Unfortunately, her son did not want to be singled out and therefore did not attend the field trips. With regard to her younger son, the school did compromise with her. They kept the AED in a knapsack with the teacher so her son would not appear to be different. She also shared that she did not want the sight of the AED to be a constant reminder to her son of his affliction.

Tori talked about petitioning the state to put AEDs in the schools within her district. She basically stated the school was very cooperative and supported her advocacy for the funding of the defibrillator. Additionally, Gloria talked about going to school at the beginning of the year and educating staff and teachers about her son’s condition. She reflected that when her son was younger, the school nurse would talk to and educate his peers about long QT syndrome. She reported this experience as supportive.

Sports, Field Trips, and Vacations

All mothers, or 100%, expressed concern regarding sports activities, field trips, vacations, and their children’s overnight stays with peers. Several of the mothers expressed disappointment about the restrictions of their children not being able to participate in sports. Lucy believed it was “just the child’s basic right to run around.” Betsy talked about needing to redirect her children to join a bowling team instead of participating in other sports. Christy expressed deep concern that her son would have “lifelong restrictions primarily with the competitive sports.” Another mother expressed that her son’s friends all play soccer, but that the physician wanted her to find another outlet for the child. Gloria stated, “and it is very restrictive as to what he can do because he loves sports. He wants to join a team. But he cannot and that really bothered him.” Mattie was concerned that her son would miss out on sports, which she perceived as a primary socialization process. Lily talked about how guilty she felt that she had to pull her son off of the sports team when he was in high school. Tori’s family revolved their life around sports. And as mentioned earlier, Mary felt deeply about her son not being active in sports.

For those mothers whose lives were not so involved with sports, the idea of sleep overs, vacations, and field trips were of concern. Betsy talked about the struggle as to whether or not to take a defibrillator on vacation. Another mother reported that her son wanted to go to a haunted house on Halloween. This was a conflicting situation for her and her husband. They also have concerns about her son going to an amusement park and which particular rides he can go on without inducing cardiac incidents. Mattie would not go on an overnight trip with her family “due to the fear of the medicine not maintaining a constant temperature.”

Family

Immediate and extended family was another area of concern for some of the mothers. Two of mothers had difficulty with their families of origin. Christy stated that "family members wanted to treat it like a cold." She talked about how difficult it was and is for her siblings and parents to understand what LQTS is and how serious it is. Mary stated that it was a great struggle to convince her mother to be tested for LQTS. Finally, after pressuring her mother, she was tested. It was soon found that other members of the family had LQTS. With regard to her spouse, Mary stated that she and her husband addressed and dealt with LQTS differently. She stated that she liked to talk about it, but that her husband did not, although she did state that he independently did do quite a bit of research about it.

Sally reported having great difficulty with her in-laws. She felt that the diagnosis in her newborn opened old wounds within the family system. Information began to come out about her brother-in-law's death several years ago. Sally believed that her brother-in-law may have died from drug-induced long QT. She stated that her in-laws did not want to talk about LQTS. In fact, she stated, ". . . and I feel judged by them." She continued, "Where is the support from the family?"

The other mothers in the study did not reveal much conflict with other family members. Most of the mothers appeared to have an intact family system. While some of the individual family members and extended family members were not supportive, all of the mothers appeared to have a supportive spouse.

The Children and Developmental Issues

All of the mothers expressed navigating through various challenges with their children who have LQTS. These ranged from communication problems, developmental issues, and medical compliance to instilling self responsibility and self-care in their children. In addition, all 10 mothers, or 100%, expressed some concern about their children's future once they left the nest. The mothers' biggest fear appeared to be not having the ability to monitor or control the children's immediate environment once the children are independent. Unlike the typical mother who may have her own separation anxiety or "empty nest syndrome" when her children leave home, the mothers of LQTS children seemed to communicate a sense of dread or distress about their children's physical well-being and medical compliance while living independently.

Explaining LQTS and Medical Compliance

Several of the mothers expressed concern about explaining LQTS to their children. Mary, Christy, Sally, and Nancy, all of whom had young children, struggled with when and how to explain the full ramifications and consequences of LQTS to their children. "The challenge for me is going to be finding that fine line between explaining to him that this is what you have yet letting him know he can have a healthy normal life. I do not want this to affect him" (Christy). Mary recounted her son's lack of full comprehension. "There have been times when he said something like 'Am I going to die?' You know, it is sad. And of course, we say 'no. You take this medicine.' He knows that there is something wrong with his heart in simple terms." Nancy also addressed her son's developmental incapacity to comprehend the LQTS. She talked about the thrill rides that her son wants to go on and her conflict of saying no and explaining to him why he is not permitted without instilling undue fear. Finally, Sally shared that her son didn't fully understand and that she remembered her son saying to her, "Mom. what would happen to me? Like, could I die?" Sally stated, "That's the hard part to deal with."

Lucy, Gloria, and Mary had young adolescents with LQTS. Their children's adolescent developmental issues appear to overrule their comprehension of the full consequences of their illness. These mothers contended with communication problems, while the children struggled with issues of identity, peer pressure, and independence. Lucy talked about trying to explain the concept of LQTS to her son. "He only just this year learned that this was called sudden arrhythmic death (he is now 14 years old). We kept that from him. He understood he was not allowed to get his heart rate up. We did not go into details. But now he is old enough to understand. You don't want to scare the shit out of a 9-year-old by saying if you do something you're going to die." She also shared that he had reached at the age at which he had a friend with a car. Lucy was concerned that her son will not listen to her and that he will begin to experiment with recreational drugs and alcohol.

Gloria also had distress about her son's noncompliance. In fact, Gloria and her mother both wanted her son to have an internal defibrillator implanted because of her son's physical activities, getting into fights, and his rebelliousness. "He is very determined. He wants to do things. He likes to ski."

Mary, who had both a young child and a 15-year-old son, had a difficult time communicating with her 15-year-old son and gaining control over his medical compliance. “That is a huge challenge. You know, he is just not open. He is not ready to talk about it. I mean I would just love to sit and explain to him, you know. But he just builds up a wall. It is a challenge.” Mary also suggested that her son had become more rebellious since having found out about the diagnosis of LQTS. “But he has become a little bit defiant. You know, once he learned he had this, he wants to challenge you, you know, like things he would never have done before. Not even the type of personality he is. He would never be defiant. But he would like, just start bike riding and going as fast as he can, you know, all this kind of stuff. Just trying, I guess, to test it.”

Lily, whose son was 19 years old and away at college, described worrying about her son’s medical compliance. She expressed a communication problem between the two of them. She stated that her son did not listen to her and that all she could do was nag. She also suspected that her son was overdoing it with exercise, over-the-counter medications, and caffeine. It was also at this point that Lily shared that she felt terrible because she had an implanted defibrillator and her son did not. She stated that she would like her son to have an implanted defibrillator because she felt like she was taking care of herself and not her son. Lily did not state whether she discussed with the physician the possibility of having her son obtain an implanted defibrillator. She also was not clear as to whether her son would even want to have the medical procedure.

Instilling Responsibility and Self-Care

Some of the mothers were concerned about how to instill self-care and self-responsibility in their children regarding the disease. They contemplated how developmental issues would affect their communication and relationships with their children, thereby affecting their children’s willingness to comply medically. Some of the mothers addressed the importance of talking and educating their children about the seriousness of their chronic illness. Lucy and Betsy both talked about continually educating their children. They both contemplated how to teach their children about limitations and how to encourage them to become their own advocates. Christy talked about trying to her “fullest to tell Bobby the truth” when he is of age. She stated that she did not want him to “live in a bubble.” She wanted him to feel normal, but also wanted him to know the seriousness of the condition and how to act responsibly. Nancy talked about giving her child “guidelines to live by.” She also talked about instilling the importance of not experimenting with drugs and alcohol as he comes of age.

Several of the mothers seemed to imply that they lost the battle. For example, Gloria’s hoped to convince her son to get an implanted defibrillator, since he was not behaving responsibly and following the medical protocol. Actually, during the interview, Gloria discussed the possibility of pressuring him into the procedure before he is 18 years old. Their hope was that they still have the legal and parental authority to do so before he is legally an adult. Lily also yearned for the same. She hoped that her son would have a nonfatal incident which would result in an implanted defibrillator. Mary talked about having faith that her younger son would be adjusted to LQTS and be more compliant because it was diagnosed at a young age. With her adolescent son, her battle of imparting self-care and self-responsibility was ongoing.

Reemergence of Uncertainty: Leaving the Nest

All of the mothers expressed worry and concern about their children leaving home for college or establishing their own independence. “I’m fearful for him when he goes to college. I will have worries about that. So, is he old enough to be able to manage his life like I am able to manage mine? I want to believe that if we take our medications and we follow the rules we will have a long healthy life” (Lucy).

Another mother returned to her 9/11 analogy. “I worry that they will stop taking their medicine or become lackadaisical about it. I worry they will decide they don’t need their medicine anymore or they will decide that when they get older that they don’t have to continue to see the doctor. There are just some things, like in general, them not bothering about the situation. I do worry a little bit about the 9/11 effect of any incidents in their lives by lulling them into a false sense of security.” Christy reported, “I cannot lie and say that the future will ever go away. I’m always concerned about the future. As I just said, it will get more complicated as he gets older and independent and will leave mom and I won’t always be there to protect him.”

Nancy shared that she worried about when it is time for her son to go to college. In fact, she stated she will even worry about him once he attends high school. She questioned whether he would remain compliant with his medication as he gets older. Sally stated, “it gets harder as he gets older.”

Gloria, because she was already having difficulties reinforcing her son’s medical compliance, stated “I just worry about him on his own. I worry about him because he denies it. He refuses to believe that he has a serious heart condition that could be fatal. The more you stress on him the more he rebels. He does not want to hear."

Mattie, who contended with her own LQTS through most of her adolescence and adult life, worried about the future, believing that the older her children get, the more likely it is that an incident will occur. As mentioned previously, Lily's son was already in college and her greatest fear was being realized as she relayed, "I worry. He wants to climb mountains. He can't climb mountains. Just like more fun things. You know, I worry about something bad happening. I actually, as much as I hate to say it, I probably would feel better if he had one" (pointing to her implanted defibrillator).

Tori reported, "I'm a little concerned that Timmy will go to college next year and, if you know, he is very good taking his medication. Obviously, nobody's perfect, especially teenage boys. Sometimes they forget. I'm glad he went to the college down the road. It is very close to us. So I can monitor him a little bit more. You know, to check with and make sure he is taking his medication."

Four of the mothers were also concerned about their children passing the gene onto the next generation. Referring to her children with LQTS, Betsy stated, "maybe they won't want to find out with their own kids because maybe this is something that they will wish they didn't know they had." Her concern was that her sons would just go into a state of denial and not follow through with the appropriate medical care of her grandchildren.

Christy and Mary both expressed concern about the gene being passed down to the next generation. Tori contemplated the same concern by wondering whether her son and daughter will marry someone else with the same genetic mutation, which she worried would increase the chances of her grandchildren having it.

Coping

Medical Compliance

Mothers were asked how they coped with their child's long QT syndrome. As noted earlier under Behavioral and Fear Management, all of the mothers used medical compliance and attempts to alter the environment as a means of gaining control over the disease. Mothers talked about feeling comforted in knowing that their children were on medication. Other mothers were comforted by believing that following the medical regimens, such as restricting diet and physical activity, gave them a semblance of control. A number of the mothers took comfort in knowing they had an AED at their disposal. Clearly, the mothers felt their best odds were to maintain medical compliance.

Support

Five of the mothers referred to family and social support as a means of coping. Lucy talked about "talking to other moms" who have children with LQTS as a means of helping her to cope. Christy talked about reaching out to other people who were affected by LQTS. However, she did state that there is a "catch 22." What she meant was that it was difficult for her to be around other mothers or families who have lost children as the result of LQTS. She was clear that she only wanted to be around other mothers who had a positive attitude. In addition, Christy was the only one who revealed that she sought professional psychological support. Nancy talked about getting to know other mothers through the hospital. Sally talked about the importance of her husband being a support to her. Gloria relied on her mother and friends.

9/11: The Passage of Time

Six of the mothers talked about the passage of time as a means of desensitizing from the anxiety and coping with the uncertainty as it relates to LQTS. Betsy, who used the analogy of 9/11 when relating her experience, suggested that being desensitized from the fear and anxiety over time is a double-edged sword. “When I mentioned 9/11, I just meant that the anxiety about it is gone. The anxiety about the kids being out of my sight or being off doing something or whatever, that part is gone. Mentally, the longer time goes by and everything is fine, the more you’re able to day by day put it aside as the occupying thought. The longer time goes by when another incident doesn’t happen, the safer you begin to feel again. Now, you know, that doesn’t mean that anything is actually changed.” Therefore, for Betsy the passage of time eased her anxiety. However, in the back of her mind she knew that at any given time, one of her sons could have a serious incident, possibly leading to sudden death.

Lucy talked about how she was “more freaked out” when she first heard about it. She stated that it took her several months to incorporate it into her life. However, for Lucy, she had a belief that her son would be protected as long as he complied with the medication and restricted his diet and physical activity. Nancy expressed gratitude for her physician. She stated that the doctor was correct when the physician stated, “‘You will get used to it in time.’ It was true. She was right. I sort of think that with time we accepted it. Like in the beginning, it was a death sentence kind of thing.”

Gloria addressed the mass confusion of first learning about LQTS and her son. “In the beginning, I didn’t know how it was going to impact his life. Now that we lived with it, and you know he’s made it all this time” things have become more tolerable. Tori reflected, “I guess it has become less dramatic. I don’t want to say less important, but after you live with it for the eight years, precautions can be taken. It is very treatable.” Finally, Nancy went on to say, “a bit more time has passed and there have not been any incidences. So, I guess I am a little bit more comfortable with it.”

All of these mothers referred to the importance of time. Each of them talked about how time has assisted them in reducing their anxiety. It should not be confused with the idea that time has erased all of their anxiety or worry about the unpredictability about the illness. However, according to these mothers, it has certainly acted as a medicinal salve.

Education/Knowledge

Six of the 10 mothers, or 60%, referred to educating themselves and others as an important aspect of their coping with LQTS. Nancy became a CPR instructor and petitioned schools to install AED. Christy talked about the importance of asking questions and educating herself in order to cope with the disease. “By understanding and knowing the disease, I know what I am up against.” Mattie was very clear about the importance of self education. “Education is the key. Understanding, for me, in order to cope and comprehend is everything.” Tori supported this concept when she was asked about what helped her to cope: “To do more research.” Finally, Mary talked about coping in terms of, “I guess just try to learn about it. Just to educate ourselves about it, you know.” Sally also referred to the importance of understanding it and learning about it.

Spirituality/Religiosity

Fifty percent of the mothers referred to religion or spirituality as a way to cope with LQTS. “It is just that it is in God’s hands. I felt like we knew for a reason so we could take the steps” stated Betsy. Christy “thanks God” about finding out about the LQTS. She also mentioned that she did not believe in coincidences. As a result of going to an LQTS seminar at a university, she related how they met “a nice couple” who gave them a name of a physician whom they stated was “the best in the industry.” For her, this was no coincidence. It was fate.

Nancy talked about surrendering her powerlessness over LQTS to a higher power. “God, or whatever higher power that you believe in, you know, has to take care of the rest. And you know, whatever is meant to be . . . you know, I think I’ve done everything I can with covering the bases at his school, the teachers, and the defibrillator. I mean there is a spiritual aspect to it. I had to look inward and outward and above. Yes, but there was definite religious or spiritual aspect to me coming to that acceptance.” Sally talked about having “a strong faith.” Finally, Mattie affirmed the other mothers’ reliance upon faith by stating “my faith has helped me a lot. My faith, I am a firm believer that God has a plan and no matter what is wrong with you, your time will come.”

Cognitive Techniques

A number of mothers also described a combination of cognitive techniques to cope with their children’s LQTS. The mothers used a combination of thought-stopping processes, cognitive reframing, minimizing, and rationalizing as a means to cope with their children’s LQTS. For example, Lucy stated, “I don’t think about it that much. I don’t think he is as vulnerable as they think. He’s hardier than they think. I think he only has a mild case of it. Look at me. I did everything that I was not supposed to do and I’m okay.” Christy exclaimed, “I just tell myself that I need to get a grip on this because, it is not a productive way of living my life by worrying about this and thinking about this all the time. I get to the point where I had enough.” Nancy unknowingly affirmed the thought-stopping and self-talk process by stating, “I sort of talk myself through it. I try not to think about it. I try not to talk about it all the time because I personally find that difficult.”

Sally explained a type of reframing as well as a thought-stopping process. “I was thinking, you know, he is going to be okay. And I think you have to do that. If you dwell on it, I would never leave the house.” Sally also reframed her challenge into a sense of gratitude by stating, “If he had died, there is not anything you could have done about it. At least now he has a chance at life. This is how I look at it. I know it is hard. But that is what I thought.”

Lily attempted to reframe her son’s illness by stating, “I just tell myself that it is better than having something where you are sick all the time. I mean it could be worse.” She also employed thought stopping by stating, “You just put it in the back of your mind and take the medicine and act responsibly.” As with Lucy, Lily also incorporated a bit of rationalization, denial, and minimizing. “I don’t believe it. I don’t buy that it is as deadly as they say.” However, later in the interview, she confessed that she knew of somebody who has a child who just died of LQTS. Lily appeared conflicted and appeared not to be coping optimally with LQTS.

Tori also used cognitive reframing as a method of coping. “We were just so thankful. You know, most occurrences are death at the time you find out about it. We are thankful, obviously, that that didn’t happen that day. Obviously, I feel we have been given a gift by him being diagnosed.” It is also apparent that the mothers who used spirituality and religion as a method of coping tended to use the reframing method to turn their difficult circumstances into a sense of gratitude.

Social Problem Solving and Problem Orientation

Most of the mothers appeared to use or attempted to use a rational problem-solving style. For example, several of the mothers addressed parceling harder concepts and medical requirements into simpler and attainable tasks. This problem-solving style also appeared to be associated with discussing and generating various alternatives regarding sports, field trips, and overnight stays with physicians and other family members. These mothers also created alternative activities which assisted their children’s quality of life, such as becoming a soccer goalie instead of a field player or playing golf instead of other, strenuous sports. After trying various alternative activities, which may have lacked appropriate accommodations for inhibiting cardiac triggers, most mothers explored and generated additional options.

Positive problem orientation was also displayed by several of the mothers’ cognitive reframing processes of their children’s LQTS affliction. The mothers who incorporated spirituality or religiosity as a means to cope tended to display a positive orientation and approach to their perceptions of the illness. Instead of perceiving LQTS as a lifelong burden, these mothers chose to or attempted to reorient their attitude to gratitude. This is not to say that these mothers did or do not contend with anxiety and stress on a daily or periodic basis. However, these mothers did attempt, although not always successfully, to face their children's life-threatening challenge by creating a variety of solutions that would enhance their children's lives, instead of perseverating on negativity and despair.

By comparing the problem-solving styles with the data which emerged from the interviews, three of the mothers, although attempting to use a rational problem-solving style, tended to fall a bit more short than the other women. For these three, their rational problem-solving style appeared to be disrupted by intermittent avoidant and impulsive/careless problem-solving styles and behaviors. For example, one mother tended to use a more avoidant problem-solving style when it came to coping with her own LQTS. From the interviewer's observations and the data, she simply pretended it is not serious. Two other mothers tended to incorporate an impulsive/careless problem-solving style when attempting to deal with their children's long QT. Instead of maintaining consistency with the disciplining of their children's medical compliance they tended to vacillate between giving into their children's demands and later attempting to reestablish parental boundaries and reasoning. This is not to say that these two mothers were devoid of a rational problem-solving skill set. There is evidence that they attempted to reinforce the children's medical compliance. However, it is likely that their anxiety, frustration, and fear impeded or curtailed their reasoning and disrupted more adequate parenting skills.

Along with the more avoidant and impulsive/careless problem-solving styles, as evidenced by D'Zurilla and Nezu's (2007) research, these women tended to display a stronger negative problem orientation towards LQTS. It appears these women were not able to shift to a more positive problem orientation. They tended to remain in or vacillate between denial and defiance or anger when talking about the disease.

Resources That Assisted With Coping

The mothers were asked which resources they found most helpful in dealing and coping with LQTS. Seven of the mothers described the importance of the medical community in assisting them with understanding LQTS. Lucy and Nancy both stated that the nurses were a great support to them in contending and coping with LQTS. Five other mothers, including Lucy, spoke of the importance of the availability of the doctor to discuss their concerns regarding their children’s LQTS.

Eight of the 10 mothers emphasized the importance of having access to various websites. Many of the mothers mentioned Sudden Arrhythmia Death Syndromes Foundation (SADS) and Cardiac Arrhythmias Research and Education Foundation (C.A.R.E. Foundation). Only a few of the mothers were aware of the C.A.R.E. Foundation. Even though many of the mothers expressed the importance of the websites, a few of the mothers found the websites to be a mixed blessing. Some of the mothers talked about not being able to tolerate the sad stories of other individuals losing their children to LQTS. For example, Nancy, Betsy, and Christy were a few of the mothers who were grateful about having the websites. They also shared that some of the stories were too sad for them to tolerate. Therefore, they used the websites less frequently. Three of the mothers participated in conferences. All three reported having positive experiences by attending conferences and felt that they gained support and understanding.

Mothers' Understanding of LQTS

All 10 mothers felt that they had some understanding of LQTS. For example, Lucy stated, “I understand it as much as I can as a layperson.” Betsy stated that she made a point to educate herself. Nancy and Mattie, both of whom were nurses, believed they had a reasonable understanding of LQTS from a medical perspective. Gloria addressed the concept that she “had to learn,” due to all the medical confusion with regard to her own misdiagnoses. The rest of the mothers all said that they understood it to varying degrees.

However, it should be noted that although the mothers believed that they understood LQTS, some of them remained a bit confused regarding the actual risks and medical understanding. For example, Lucy did not think that her son had a major case of LQTS, whereas Sally simply stated that sometimes she thought she “doesn’t understand it” and that she felt the doctor’s understanding is “way above” hers. Finally, Mary talked about understanding it to varying degrees.

Mothers' Experiences and the Medical Community

At the end of the each interview, the mothers were asked if there was anything that they wanted to share that was not covered during the interview. Five of the mothers volunteered that they felt there were problems within the medical profession regarding LQTS. Lucy recounted her experience of indifference from the emergency room doctor and pediatrician after her son's fall. Betsy said that she received conflicting advice from different physicians. Although not reporting specific details, she felt some of the doctors were using invasive measures that were inappropriate. She stated there is a "big difference having doctors who use invasive measures and those who don't. Parents who were put in this position should be made aware of having different opinions. Parents should not be just influenced by a particular opinion of the doctor who happens to be treating their kid." Betsy continued, "I think a lot of times the parents don't get the information. I think that is why people turn to the Internet for things. They try to find it on their own. People don't trust that they are getting the whole picture. I think if doctors made more of an effort to say, 'this is what my experience tells me' and would say that there are other people who see it differently, and 'I will recommend this, but others can recommend something else.'" She went on to state, "I feel like there are probably things that people are finding [referring to medical profession] out that are not being disseminated. It would disappoint me to find out that people were doing a lot of things, you know, everything from genetic testing to more accurately diagnosing the disease." Betsy appeared to have developed a general mistrust of the medical community.

Christy's biggest concern was that she felt that the medical community did not take into account her psychological well-being upon hearing the news. "I wished that the medical industry could understand the psychological side effects of a parent being told that your child has this condition." She felt "that the doctor does not understand the psychological effects . . . . "

Mary talked about reading stories about how doctors have underdiagnosed LQTS. Her concern, along with a number of the other mothers, was that there needs be a stronger public awareness of this illness. Gloria confirmed this by sharing about her admissions to the emergency rooms and how her syncope and seizures were misdiagnosed.

Although five mothers initially had difficult experiences, most of them overcame their negative experiences and found comfort in their medical care. Although it may appear to be a contradiction, eventually there were at least seven mothers who found solace through health care workers and physicians in spite of initial problems. It should be noted that there is an overlap with the mothers. For example, even though Lucy initially had a difficult experience, she later stated that speaking to the doctors and the nurses had been a great support for her. Christy also reported finding comfort in her child’s physician, although she believed that appropriate transitional services from first learning of the diagnosis to living with it on a daily basis were not adequately provided. Finally, four other mothers mentioned finding solace from their physicians or the hospital staff. Another mother also mentioned that interacting with the general hospital staff was a positive experience for her. However, it is noted that the more positive experiences appear to develop as a result of an ongoing relationship with the physician and the clinical staff.

Insurance

Finally, three of the mothers shared their frustration with the insurance companies. Betsy talked about the cost of genetic testing and the lack of affordability. She also stated that they had to fight the insurance companies for the testing. In addition, she expressed concern about her children's ability to obtain good health insurance when they become adults with a preexisting condition of LQTS.

Nancy stated, "Well, I guess the thing I really can touch upon, this frustration with health care . . . It was really frustrating. We were fighting with the insurance company." Nancy stated that it took several years and numerous appeals in order to cover their cost of the genetic testing.

Summary of Findings

During the interviews, the mothers were open and responsive about their children's and their own (where applicable) experiences with LQTS. Each mother had her own unique experience as to how she first learned and became aware of LQTS in her family and children's lives. The salient features which emerged from these interviews included how each mother attempted to assimilate and cope with an unfamiliar life-threatening and chronic illness in their children. Each mother had the task of comprehending and integrating large amounts of medical information within a short span of time. In addition, as data emerged, the types of effective cognitive-behavioral processes related to social problem solving and coping were identified, while the less effective problem-solving skills were noted. Most importantly, unique insight and knowledge into living with LQTS on a daily basis was revealed by the mothers' personal and intimate stories, thereby exposing aspects of their daily challenges, how they coped with those challenges, and a perspective regarding the quality of their lives.

CHAPTER 5

Discussion

Theoretical Analysis of Descriptive Findings

All the mothers involved in the study had undergone a transformative experience as a result of either learning they had LQTS and/ learning their children had it. This journey began with the monumental task of comprehending and assimilating the biological aspects of long QT. For example, in Cohen's (1993a) study interviewing parents of children with chronic illnesses, she suggests that not only is the news of the child's life-threatening illness shattering for the parent, but the parent must rapidly assimilate large quantities of highly complex medical information which she (1993a) describes as "generally conveyed in ambiguous language" (p. 141). This was found to be true within this study as well. However, the highly medical intricacies of long QT syndrome and the complexity of it only served to exacerbate the mothers' distress while they attempted to assimilate all aspects of the illness, including the biological, psychological, and social implications. As a result, all of the mothers demonstrated a multidimensional experience of accommodating themselves to their own (where it applies) and especially to their children's chronic illness.

The mothers' experiences support a concept in health psychology known as the biopsychosocial model. The biopsychosocial model is a holistic approach of viewing individuals' experiences in a multidimensional world. The core foundation of the biopsychosocial model is for health practitioners and psychologists to integrate the biological, psychological, and social-cultural milieu of the individual (Belar, 2003) in order to prognosticate, assess, and promote physical, social, psychological, and emotional well-being. As demonstrated within the discussion of findings, each of the mothers initially needed to contend with and assimilate the medical/biological understanding and the consequences of long QT syndrome. In addition, they needed to cope with their psychological and emotional reactions. Finally, each one of the mothers had their own particular social milieu in which they needed to incorporate the new health ramifications of their children’s chronic illness.

In summation, the findings within this study can be broken down into five primary stages paralleling a biopsychosocial model. Stage 1 is biological assimilation and the impact of incorporating and comprehending LQTS. This stage includes the mothers’ initial attempt at assimilating and adapting to the biological understanding and consequences of LQTS. Stage 2 incorporates examining the mothers’ initial psychological and emotional reactions to the diagnosis. Stage 3 includes the aftereffects of the initial reaction of the diagnosis and the evolving psychoemotional states. Stage 4 is the mothers’ attempts at integrating LQTS into their social milieu. Finally, stage 5 is the mothers’ reemergence of uncertainty as they face the prospect of their children’s futures. It should be noted that the stage model is not linearly based. Each mother has her own particular set of circumstances and experiences. On any given day, it is possible for a mother to revert back to fear, sadness, or even confusion about the understanding of LQTS. The next day, a mother could be surrounded by positive circumstances and feel happy and secure in her life and her family’s well-being. As with any parent/child relationship, the relationship is dynamic, not static. As the parent/child relationship evolves over time, especially related to developmental factors, it is likely that the mother may enter any one of the stages at any time, given her evolving perspective.

Stage 1. Biological Assimilation: The Impact of Incorporating and Comprehending LQTS

Factors Affecting Mothers' Reaction to Diagnosis

Sudden onset. Brown (2003) found in her study of children with congenital heart disease that the suddenness of onset of the disease and its unexpectedness contributed to the distress upon parents' first hearing of the diagnosis. Seven of the mothers from this study appeared to have been directly affected by the suddenness of the symptoms and diagnosis of LQTS. Research suggests that a parent experiences more distress with a sudden onset (Dodgson et al., 2000; Hauenstein, 1990), which has been affirmed by most of the mothers in this study. For Lucy, Tori, and Mary, the news came suddenly and shockingly and was initially and solely related to their children, whereas for Christy and Lily, it was the sudden onset of their own condition. With Betsy, it was her husband's cardiac arrest which prompted her initial distress. Shortly thereafter, for all the mothers, the news of the diagnosis in their children added to their shock and anxiety.

Mattie, on the other hand, experienced her own sudden onset when she was 14 years old. Additionally, she had to face the possibility of her children being born with LQTS. Unfortunately for Mattie and her children, she "lost the lottery." Nancy did not appear to be too affected by the news of her own LQTS. However, the suddenness of her child's symptoms appears to have been a primary factor in her distress.

There appears to be a break from Brown’s (2003) findings regarding Gloria and Sally’s experience. Both Gloria and Sally appear to have had a somewhat delayed reaction. Sally was told soon after the birth of her son, but her distress did not seem to appear until later, when her knowledge about LQTS evolved. Similarly, Gloria appeared not to take her diagnosis too seriously initially because there were no clear symptoms. However, several years later, she experienced the sudden onset of her own symptoms. In addition, she had numerous trips to local emergency rooms due to seizures and syncope from the LQTS. Disturbingly, according to Gloria, it was not until a year later that her condition was accurately diagnosed, at which time her symptoms were brought under control. After experiencing her own severe symptoms and the misinformation regarding LQTS, she became much more distressed about the illness. It was also at this time that she appeared to transfer her anxiety about her experiences onto her son.

Disease type, severity, and prognosis. Hauenstein’s (1990) and Board and Ryan-Wenger’s (2000) studies conclude that the type of illness itself in divergent chronic conditions will generate different parental stressors. Closely related to disease type and prognosis, Board and Ryan-Wenger (2000) suggest that mothers’ perception of the severity of the illness is an additional factor affecting mothers. In a more recent study specific to LQTS, Hendriks, Grosfeld, Tintelen, van Langen, Wilde, van den Bout, and Kroode (2005) administered questionnaires to 36 parents whose children were recently genetically tested for long QT syndrome. Seventeen of the parents were found to have children carrying the gene for LQTS. They found that the parents of children with the carrier gene did not adjust as favorably as the parents of children without the gene. They suggest that parents who have children with the LQTS gene have higher levels of distress because of the possibility of the sudden development of potential fatal symptoms. In fact, they state that 33% of the parents with the children carrying the LQTS gene in the study maintained a strong preoccupation with the disease at an 18-month follow-up. This likely implies that a significant number of parents with children with LQTS perceive a stronger severity associated with LQTS than possibly with other chronic illnesses. Although it should be noted that the mothers in this study, for the most part, had adjusted to living with their children's LQTS, all of the mothers had at least some level of ongoing concern in monitoring behavior regarding their children's disease. This is congruent with the findings of Hendriks et al. (2005); the parents in their study did not adjust favorably, which was likely due to the perception of the potentiality of sudden and fatal symptoms from LQTS.

Etiology. Although the etiology of LQTS is somewhat understood from a medical perspective, it remained ambiguous and daunting to many of the parents in this study. For some of the mothers without a medical background, LQTS appeared to remain a vague and abstract concept. Each mother admitted to having varying levels of understanding. A few of the mothers continued to have various misconceptions and misunderstandings of the disease. For example, one mother believed that as long as her child takes medication, he will be protected. Another mother believed that the disease is not as severe as the medical community makes it out to be. One of the mothers expressed continued confusion about the disease, even though significant time had passed since the condition was diagnosed in her child. She simply stated that she just did not understand the mechanics and why or how it happens. What is interesting is that those mothers who were diagnosed with LQTS prior to their children tended not to describe the same heightened distress upon learning of their own LQTS as when they learned of it in their children. For example, Hendriks et al. (2005) found that parents were more focused and concerned about their children’s well-being than their own. In addition, the *DSM-IV-TR* reports that health concerns of a family member or child will affect psychological well-being (2000). All said, it is likely that the mothers’ perception of their role as protector of their children has been undermined by the very nature of LQTS, leaving them without the ability or guarantee to protect their children from the possibility of a sudden cardiac incident.

Other Factors Unique to This Study

Witnessing their child’s and/or having their own LQTS incident. The mothers who initially reported the highest level of distress were Lucy, Christy, Betsy, Gloria, and Mary. The factors that these mothers had in common were either having experienced a significant incident or episode of their own or the witnessing their child’s incident. Although the witnessing of symptoms was not discovered in the review of literature, it does appear to be a factor in this study. All these mothers reported a shocking, devastating, or fearful experience upon witnessing an episode and learning of their children’s LQTS, in addition to their own episode.

Mary, who had no prior knowledge of LQTS, spoke in horror of witnessing her son’s unconscious state. Although Nancy was a nurse and previously knew she had long QT syndrome, witnessing her son’s likely syncope and loss of consciousness increased her distress level. Lucy did not actually witness her son’s fall. It was the consequences and physical injury from the falls, in addition to the unfamiliarity of LQTS, that escalated Lucy’s distress level.

In Christy's case, she was pregnant with her first son when she learned of her own diagnosis of LQTS. Because Christy had her own serious symptoms of LQTS, such as syncope or a cardiac incident while she was pregnant, she maintained a great concern for the impending birth of her child. When her son was born and LQTS was diagnosed shortly thereafter, her distress level increased. In fact, she stated that it initially felt like it was a "death sentence" for her son.

Gloria's increased disturbance was developed through her own series of syncope, seizures, and cardiac incidents. While Betsy witnessed her husband's cardiac episode, her subdued distress about her children became obvious when comparing their incidents to 9/11.

Medical professionals' influence upon maternal distress and the disclosing of the diagnosis. This study found that several of the mothers either initially described having a negative experience when learning of the diagnosis or perceived the medical community as lacking in appropriate knowledge regarding the disease. It should be noted that the ambiguous nature of LQTS may influence the mothers' perception of the medical community's knowledgeability. Therefore, even the most informed physicians and medical staff likely struggle to communicate the complexity and uncertainties of this disease.

Mothers perceived that having a difficult or contentious experience with medical professionals only perpetuated their anxiety, fear, and distress, thereby hindering comprehension of the illness. For example, one of the mother's experiences with the ER doctor and other medical professionals tended to add confusion and stress to her understanding of LQTS. She appeared to still harbor some misinformation. Another mother seemed to have directed her frustration and misconceptions towards the medical community. She believed, at best, that the doctors are ambiguous about the diagnosis and indefinite about treatment of LQTS itself. From that experience, as well as her internal conflict, she appeared to maintain a state of masked anxiety and frustration. Gloria, of course, had serious cardiac episodes, seizures, and syncope. After numerous trips to emergency rooms and her cardiologist's inability to appropriately diagnose the symptoms, Gloria remained frustrated by the occurrences. In addition, these experiences have only compounded her anxiety about her son. Mary, although she was grateful for having some of the best doctors in the field, referred to her heightened level of anxiety due to learning of the diagnosis of her son's LQTS from what she perceived was indifferent medical personnel. For example, she was upset that son was discharged from the hospital on a weekend after his cardiac episode with "only a prescription" for beta-blockers. She described feeling exiled to care for a child with an uncommon disease with the only advice being to call the cardiologist the following Monday. Finally, Christy, who placed her faith in the medical professionals as well, strongly believed that her anxiety and distress were multiplied due to her introduction to the diagnosis and a lack of psychological understanding on the part of the health care professionals. Christy stated that she would like physicians and medical staff to understand and explain the emotional complexity of what a mother may experience when learning about her child's LQTS. She shared that she was only counseled in the medical symptoms and not given a guideline as to what the psychological side effects could be.

For the other mothers, the concept of the diagnosis and symptoms was emotionally staggering. However, the other mothers had a more positive and supportive introductory experience with the diagnosis through the medical milieu, which likely eased the initial distress or trauma.

Summary of Biological Factors

Due to the limits of this study and the small group of subjects, it is impossible to specifically determine what combination of factors leads to higher levels of distress. However, it is possible to identify general patterns suggesting which amalgamation of factors may lead to higher levels of stress and anxiety. None of the mothers' initial shock and anxiety should be discounted. All mothers reported difficulty receiving the news that their children were afflicted with LQTS. This discussion is regarding a matter of degree of distress, not the absence thereof.

If the supporting literature is correct and patterns found within the study were adequate, it would be reasonable to expect that the mothers who had a combination of two or more factors would have higher levels of distress. For example, mothers who witnessed their children having an incident prior to the diagnosis or mothers who had their own anxiety-producing episode of LQTS in combination with factors of a negative medical experience would experience greater distress initially. Having little or no knowledge of LQTS in combination with one or more of the aforementioned factors would compound the mothers' level of distress upon learning of the diagnosis. Following this logic, Lucy, Christy, Gloria, Betsy, Lily, and Mary would likely be the mothers with high initial distress. Upon reviewing the transcripts, it appears these are the six women who may have had more difficult reactions and time adjusting, as shown in Table 4. For example, each was found to have experienced at least three or more of the following factors: suddenness of onset, witnessing a child or other family member having a cardiac episode, her own LQTS incident, knowledge of LQTS before it was diagnosed in the child, perception of the severity of the disease, and a difficult medical experience upon diagnosis.

Nancy, Tori, Sally, and Mattie appeared to have less distress or more moderate levels of distress than the other women. When examining the transcripts, these women experienced fewer of the factors leading to higher distress. For example, none of these women described having a difficult medical experience regarding the initial diagnosis. Sally and Mattie did not witness their children having episodes. Nancy and Mattie both had a medical background. This is not to say that these mothers did not have trauma or significant anxiety. They clearly expressed distress, but it may be surmised that the adjustment period may have been less enduring. For example, Nancy described adjusting to the traumatic news within a week, meaning the initial shock wore off. Mattie, who had years of adjusting to her own LQTS, was a nurse and knew the risks of giving birth. And for Sally, it was not until she comprehended the full ramifications that her distress level increased significantly

Table 4.

Factors Influencing Mothers' Initial Distress

Pseudonym	Sudden onset vs. diagnosed without incident	Witnessing child's or other family member's incident/episode or experiencing child's/other's hospitalization	Mother's experience of her own incident or simultaneous diagnosis	Lacked prior knowledge of etiology of LQTS before child's diagnosis	Perception of disease type, prognosis, severity	Difficult medical experience
Lucy	Yes	Yes	Yes	Yes	Moderate	Yes
Betsy	Yes	Yes	No	Yes	Moderate	Yes
Christy	Yes	Yes	Yes	Yes	Severe	No
Nancy	Yes	Yes	Yes	No	Moderate	No
Sally	No	No	No	Yes	Moderate	No
Gloria	No	Yes	Yes	Yes	Severe	Yes
Mattie	Yes	No	Yes	No	Moderate	No
Lily	Yes	No	Yes	No	Moderate	Yes
Tori	Yes	Yes	No	Yes	Moderate	No
Mary	Yes	Yes	Yes	Yes	Severe	Yes

Stage 2. Initial Psychoemotional Reactions: Denial and Disbelief

After learning of the diagnosis of their children's LQTS and attempting to assimilate the biological consequences of it, 80% of the mothers described a numbing of feelings through denial or disbelief. This follows several models regarding initial reactions after learning of a loss and in the initial stages of mourning. For example, Goodheart and Lansing (1997) discuss the intrapsychic shock when first learning of a chronic illness. They suggest that when an individual first learns of a chronic illness an intrapsychic disorganization occurs. This results in a challenge to "one's self and identity" (p. 69). Therefore, experiencing a level of disbelief, denial, or a numbing of reality would appear commonplace. This is confirmed by the classic studies of Elizabeth Kubler-Ross, who researched terminally ill patients. She asserts that the first stage upon discovering a terminal illness is denial (Kubler-Ross, 1969). This model has been expounded and expanded upon over many years to include mourning and losses that are not only related to terminal diseases, but also encompass a variety of losses, including chronic illness.

Also to be considered during this stage are the other factors discussed earlier. Depending on these factors, each mother is likely to experience varying lengths of time in each stage and/or revisit various stages at any point in time, given varying circumstances. However, it is beyond the scope of this study to determine such a timeline. For example, two of the mothers did not appear to have fully resolved the initial reaction stage of denial. They both had distressful experiences and maintained some level of disbelief and misconceptions about LQTS, although several years had passed since LQTS was diagnosed in their children.

It is also important to note that the stages proposed within this study and the Kubler-Ross stages are not linear. Depending on the individual and varying circumstances, each person may experience each stage for a longer or shorter duration. Each individual may not go through every stage or may quickly pass through one stage and revert back to a previous stage. It is also possible for a person to go through all stages to resolution, but experience an incident that returns them to a previous stage temporarily. Unfortunately, there may be individuals who remain in a certain stage of unresolved grief or adjustment and never obtain resolution (see *DSM-IV-TR* diagnosis for complicated bereavement).

Stage 3. Evolving Psychoemotional States and Coping Behavior

Emotional States and Cognitions

Fear and anxiety. After the initial reactions to the biological aspects of the diagnosis and with some time to process, the mothers had a second wave of emotions. One hundred percent of these mothers described an emergence of fear, anxiety, or continued stress after assimilating the news of the diagnosis and comprehending the ramifications of LQTS. Some of the mothers described feelings of fear, devastation, and trauma (Brown, 2003; Cohen, 1993a; Landolt et al., 2002). These emotions emerged in the mothers whether their children exhibited symptoms or not. Even if the children were asymptomatic, all of the mothers expressed a range of fear and anxiety regarding the possibility of their children's sudden death. Farnsworth, Fosyth, Haglund, and Ackerman (2006) confirm this, in part, in their study of parents who have children with LQTS. They report that of the 31 parents interviewed, 19 expressed a direct fear about their children's disease and possibility of sudden death. However, they go on to state that 12 of the parents in their study reported not having fear about the illness. The 12 parents who stated that they did not have fear had children who were asymptomatic. However, Hendriks et al. (2005) report that 75% of their parents of "carrier" children demonstrated a preoccupation and a focus on their children's symptoms, even though the children were asymptomatic and were only carriers of the gene. Hendriks et al. (2005) also suggest that the parents perceived their children as vulnerable, suggesting at least some element of ongoing fear or anxiety.

With regard to the discrepancy between this study and the studies by Farnsworth, Fosyth, Haglund, and Ackerman (2006) and Hendriks et al. (2005), it may be suggested that it is due the limited number of subjects in this study. In addition, this study was conducted in a semistructured interview format. The interview format may have provided a broader and more in-depth gathering of emotional content which may not be accessed through questionnaires, psychometric instruments, or anecdotal reviews.

Sadness and loss. Four of the mothers, or 40%, discussed feelings of loss and sadness. This seems primarily centered around restrictions placed upon their children's lifestyle. These mothers appeared to be grieving what they perceived as a loss of their children's quality of life. Several more of the mothers discussed their concerns about limiting their children's sports activities, social activities, and avoiding triggers.

Farnsworth, Fosyth, Haglund, and Ackerman (2006) also address this issue in their study, wherein they discovered that one of the most significant impacts for the families of children with LQTS was avoiding triggers, which affected the family's quality of life. It is possible that other mothers may actually have been feeling loss or sadness, but did not articulate it within the structure of this study. In addition, because this is not a linear model, mothers may experience sadness or loss due to future conditions. For example, as some of the younger children age, the mothers may once again need to face the loss of their children's engagement in more age-appropriate activities such as sports, games, or other peer-related rites of passages. Another possibility is that as the children broaden their social milieus, the mothers will once again be facing the question of whom to tell or not tell about their children's condition, in addition to reconsidering various environmental triggers. Finally, some of the mothers expressed concern that their children will have to decide whether or not to conceive children due to LQTS. This loss of not being able to perpetuate future generations could reemerge feelings of sadness and grief for some.

Guilt. Three of the mothers directly used the term *guilt* in describing their journey of coping and contending with LQTS. Other mothers implied a sense of guilt or shame. However, this seems to be more related to the mothers' concerns with whether those in their immediate social milieu would perceive their children as vulnerable or different from their peers.

Although only 3 of 10 mothers directly described feelings of guilt, the literature does support their experience. Cohen (1993a), in her work with parents of children with cardiac disorders, and Deford's (1983) experience with his child's cystic fibrosis describe how the parent would feel rather culpable in some way, thereby gaining some distorted understanding of and control over why the disease occurred.

Perpetual anxiety and preoccupation (mindfulness). Nine of the 10 mothers engaged, to varying degrees, in vigilant behavior, such as monitoring of and/or preoccupation with the diagnosis and their children's well-being. It is noted here that the term *preoccupation* may not necessary connote psychological pathology. Given the possibility of SCD at any moment, preoccupation with or "continual mindfulness" of the children's health may be a norm. This concept of continual mindfulness or preoccupation is supported by Farnsworth, Fosyth, Haglund, and Ackerman (2006), who state that 61% of parents studied remained concerned about the mortality of their children. For example, they report that it was the habit of some parents to walk into their children's rooms in the morning with a preconceived fear that they may not be able to wake their child due to sudden death. This sentiment was echoed by several of the mothers in this study when they talked about going into their children's room throughout the night or in the morning to be sure that they were still breathing.

Ninety percent of the mothers in this study demonstrated continual mindfulness of their children's vulnerability and risk of sudden death. This suggests a perpetual state of anxiety or concern, even if it is not always at the forefront of their minds. Even though not all mothers described a conscious mindfulness or perpetual anxiety, 90% of them either implied a subtle preoccupation in the "back of one's mind" or a mindfulness described by behaviors such as monitoring the children while at sleep or play or by phone calls throughout the day.

The role of cognition. It can be argued that the mothers' preoccupation with their children's vulnerability to sudden death may or may not be a cognitive distortion. As noted in the literature, an individual with too many cognitive distortions can develop intrapersonal and/or interpersonal dysfunction (Freeman, Pretzer, Fleming, & Simon, 2004; Nezu, Nezu, & Lombardo, 2004). However, since sudden death is always a possibility, even if only on a minimal scale, a perpetual concern about the children's susceptibility may not be an irrational belief, apprehension, or cognitive distortion. Unless this preoccupation or hypervigilance interferes with the mothers' interpersonal relationships, social functioning, or intrapersonal functioning, there may not be a need for concern. In all actuality, it may be reasonable. There was only one mother who appeared to exhibit an undue and ongoing distress accompanied with clear intrapersonal difficulties. However, she was in the process of remedying this by participating in various interventions.

Other mothers exhibited and expressed various cognitive distortions. The mothers who continued to contend with feelings of guilt had various cognition distortions. Three mothers expressed some type of belief that they were responsible for their children's LQTS. Not only were feelings of remorse spawned by these beliefs, but the actions which accompanied these beliefs fueled some risky behaviors. For example, one talked about her own noncompliance, her disbelief that LQTS was as deadly as it is, and how she was responsible for her son's loss in quality of life. These beliefs appeared to lead her to a state of frustration and noncompliance regarding her own medical protocol and ongoing frustration. Another mother believed that she got away with things that her child will not. Additionally, her belief that her child is "hardier than they think" (meaning the medical community) likely reinforced her denial, minimization, and wishful thinking. Finally, one expressed such guilt that it was palpable during the interview. These women's experiences are supported by other research. For example, Brown (2003). Deford (1983), and Dodgson et al. (2000) found that some parents of chronically ill children may believe that they are responsible for their children's illness, leading to feelings and cognitions of guilt.

Belief systems or negative cognitions may also affect the mothers' social interactions. A significant number of the mothers had concerns about friends', peers', and acquaintances' perceptions regarding their children's potentially fatal disease. These mothers carried the belief that the immediate community would see their children as vulnerable or different. Once again, it begs the question whether these beliefs may or may not be based on fact. For example, Tori recounted an accommodating community which embraced her children and their diagnosis of LQTS, while other mothers believed that they observed their family members, friends, and members of the community being uncomfortable with their children. Therefore, whether these thought processes can be attributed to the mothers' cognitive distortions or accurate appraisals of their community is beyond the scope of the study. It is likely a bit of both. However, it is noteworthy to address in future research.

Coping Behaviors

Fear management. As the mothers processed their feelings and grappled with the concept of their children having long QT syndrome, each engaged in reducing their children's risk level by managing their environments and eliminating, or at least reducing, environmental triggers that potentially could initiate cardiac incidents or episodes. Farnsworth, Fosyth, Haglund, and Ackerman (2006) describe this process as fear management. They state “fear is managed by parents with lifestyle changes they make in their homes and by actions they take in their communities” (p. 287). This concept is evidenced by 100% of the mothers in the study. Every one engaged in some type of environmental engineering, such as limiting physical activity, changing diet, providing cell phones, changing alarm noise levels, educating their children, and educating the community.

A number of the mothers expressed relief in gaining some sense of control by limiting and/or enhancing their children’s physical environment. The mothers also expressed faith and hope by following through with medical compliance. For a few of the mothers, their fear management was the belief that their children’s medication, such as beta-blockers, would protect them from any fatal incident, whether an accurate perception or not.

Support. As stated above, all of the mothers engaged in some structural change with the support of their spouse or other family members, as part of medical compliance, and in generating an environment that would protect their children from any cardiac symptoms. Additional support was found through relying on family members, friends, medical staff, and parents of other children with similar chronic illnesses (Brown, 2003). Five of the mothers engaged in this form of coping by relying on their spouse, going to workshops, talking to nurses/doctors, and reaching out to other mothers of children with long QT syndrome.

Passage of time. Six of the mothers cited the passage of time as a means of coping. Farnsworth, Fosyth, Haglund, and Ackerman (2006) also found that parents cope with and manage the uncertainty about their children's LQTS "with the passage of time" (p. 288). As stated by one of the mothers in this study, as time passes, one becomes more secure regarding the safety of the child. However, she also realized that there is no guarantee. With the passage of time, parents and their children begin to find solace in their daily routines. The longer the child goes without an LQTS incident, the more they take comfort in the passage of time. However, Betsy reminded herself, "the longer time goes by when another incident doesn't happen, the safer you begin to feel . . . that doesn't mean that anything is actually changed."

Knowledge and spirituality/religiosity. Six of the mothers expressed self-education and gaining appropriate knowledge as a means of coping. They found that identifying and knowing the risks, along with understanding various factors about LQTS, gives them a sense of security. In addition, 50% of the mothers referred to having faith or a strong sense of spirituality or religiosity as a means of coping. Various studies have found that factors such as hope, optimism, humor, and religiosity assisted individuals in coping with health problems such as cancer and 6 (Bremer et al., 2004; Lefcourt, 2005; Pargament & Mahoney, 2005; Peterson & DeAvila, 1995).

Social problem solving. Finally, several of the mothers engaged in some form of social problem solving (Nezu et al., 1999). Although the specific components or styles of social problem solving were not examined in detail, several of the mothers appeared to demonstrate various approaches as an attempt to deal and cope with their children's LQTS. However, what did become clearer in this study is that the mothers appeared to utilize a combination of problem solving styles. For example, one mother tended to display avoidant style of coping behavior. She readily admitted to using denial as a means to contend with her LQTS. However, regarding her child, she tended to use a more impulsive/careless problem-solving style (D’Zurilla & Nezu, 2007). For example, she talked about nagging her son about his use of over-the-counter stimulants, caffeine, and exercise, but then became self-deprecating about her reactionary behavior. She tended to go from trying to enforce limits and boundaries and using reasoning to impulsively and indiscriminately attacking him, thereby exhibiting an impulsive/careless problem-solving style that overrode her attempts at more rational problem solving.

Another mother, on the other hand, talked about breaking down into smaller tasks the medical protocol and other restrictions in order to deal with the changes. This is reflective of D’Zurilla and Nezu’s (2007) rational problem-solving style. However, this is also the same mother who tended to use disbelief and denial throughout the last few years in order to cope with her child’s illness. This does not necessarily discount her rational problem-solving style. It may simply suggest that she used an avoidant problem-solving style to distance herself from her own internal conflicts and emotions about LQTS while attempting to incorporate rational processes in dealing with her child.

Five other mothers appeared to use a rational problem-solving style. A few of them talked about volunteering at local schools to teach CPR, to talk about LQTS to peers, and putting a clear and rational plan into operation for their children. They also appeared to have clearer boundaries of authority with their children. However, even in this group, one of the mothers tended to occasionally use avoidance and denial by not taking along the AED on various trips, as dictated by the medical protocol.

Finally, the last three mothers also attempted to use a more rational problem-solving style. They all appeared to follow through on a rational and clear medical compliance regimen. However, two of them expressed difficulty with their adolescent children. It appears that the authority and structure of their relationship with their children was not quite as rational and structured, with appropriate boundaries. However, this is only speculation at best and may be considered as a topic for additional research. Finally, one mother was very dedicated to approaching and coping with her child's LQTS in a productive manner. She attempted to utilize a rational problem-solving style. However, she still was quite overwhelmed by the disease. As time goes on, and she continues with appropriate psychotherapy, it is very likely that she will develop a rational problem-solving skill set, along with positive coping skills.

With regard to problem orientation (D'Zurilla & Nezu, 2007), at least half of the mothers attempted to have a more positive outlook in coping with their children's LQTS. Fifty percent of the mothers utilized spirituality and/or religiosity as a means of coping. According to D'Zurilla and Nezu (2007), a positive problem orientation is a set of cognitive operations that disposes a person to a constructive approach to problem solving. Five mothers who incorporated spirituality and/or religiosity talked about a sense of gratitude with regard to finding out about their children's LQTS. They were thankful that they found out about the condition before a life-threatening or fatal incident occurred. By having this positive orientation, these mothers likely approached coping with LQTS in a more constructive manner. Again, three of the mothers tended a bit more to a negative problem orientation. One talked about her frustration and displeasure with her LQTS. She lamented having the implanted defibrillator, in addition to feeling guilty that her child did not have one. Another continued to minimize her son’s susceptibility to LQTS and appeared to be frustrated that her child would not be able to participate in as physical a lifestyle as she would deem appropriate. The last, on the other hand, appeared to be in transition of attempting to change from a more negative problem orientation to a more positive problem orientation.

Stage 4. Social Disarray and Reintegration

Social stigma. Seven of the mothers struggled with concerns about social stigma. Mothers were concerned about their children being perceived as different from peers or more vulnerable and susceptible physically. Some of the mothers also described a concern that if they told other individuals, they would also have to assist and support those individuals with managing their feelings as well as their own. They felt that trying to monitor or control other people’s reactions about LQTS would be a daunting task. Another concern was that their community would have misconceptions about LQTS.

For these mothers, it was not an automatic response to let their friends and those in their community know about their children’s LQTS. The mothers struggled with the question of whom to tell. Some of the mothers appeared concerned about carrying an AED with them. All of these mothers were concerned about their children being treated differently and being labeled as different. Although no mother expressed feelings of shame about their child, there is a possibility that there is an element of shame about the label of the disease. It could be analogous to a “coming out process” for the sexual minority individual. For example, most sexual minorities appear undifferentiated from their peers in their community until they self-identify. This appears similar to mothers of children with long QT syndrome. Well-meaning mothers do not want their children to be perceived as different, leading to them being possibly ostracized or treated differently. Therefore, although it would seem a natural reaction to immediately inform everyone within their children's environment, this research found that not to be so. Several of the mothers struggled with what appeared to be choosing between having their child suffer from a social stigma by informing others or taking the risk of the stigma in order to ensure their children's safety. It is not an easy choice for these mothers. Some of the mothers were likely not to inform the larger community, but only those closest to them. Only a few mothers appeared to tell the community at large.

School and family systems. Other systems addressed in this paper were the school system, sports activities, field trips, and vacations. Two mothers had difficulty with the school system, whereas two mothers found the school system to be very cooperative and supportive. Gloria and Tori both talked about the cooperativeness of the schools. Gloria referred to e-mailing and talking with the school nurses and teachers. She stated that they were very accommodating. Tori shared that she petitioned the school to obtain AED. She reported that the school was very accommodating, and they successfully obtained the appropriate devices.

Although Mary initially had a more positive experience with the school system, eventually she felt that they became too rigid in their demands. Her son stopped participating in field trips because he was too embarrassed to have a nurse stay with him throughout the trip. Lucy was vocal about her difficulty with the school. She simply stated that the school overreacted, and she believed it was just about protecting the system from lawsuits.

Eight of the mothers expressed concern about the deprivation of their children not being able to participate in sports. In addition, several of them talked about the restrictions and concerns of sleep overs, field trips, and vacations. One stated that she wouldn't even consider traveling somewhere overnight for fear that her children's medication would be affected from various temperature changes.

The family system was another area of difficulty for some of the mothers. Three mothers had difficult experiences with various family members. Each of these mothers addressed concerns that family members, apart from their children and spouses, did not want to acknowledge the seriousness of the illness.

Mother and child system: Developmental challenges. One hundred percent of the mothers described current or future concerns related to developmental challenges with their children and LQTS. Congruent with the study by Farnsworth, Fosyth, Haglund, and Ackerman (2006), the four mothers of children whose LQTS was diagnosed during adolescence appeared to have the most difficult time with their children and medical compliance. This led to additional communication problems between the mother and child, difficulty with enforcing the child's medical compliance, and, for a few of the mothers, outright defiance from their children. Finally, Tori and Betsy had concerns about long-term effects of their children's medication regimen on developmental issues (Farnsworth et al., 2006) and the effects on the body.

Similar to the findings of Farnsworth, Fosyth, Haglund, and Ackerman (2006), the mothers who had younger children with LQTS had less difficulty with their children's medical compliance. Farnsworth, Fosyth, Haglund, and Ackerman (2006) report that for those children in whom LQTS was diagnosed at a young age, it becomes more easily integrated as a norm in their children's life. In other words, the younger the children are, the more readily they adjust to the diagnosis. Several of the mothers with younger children did report this phenomenon. However, all of the mothers expressed concern about their children leaving the nest or moving on to college. Every one of the mothers expressed concern that their child may not follow through with medical compliance once they achieve independence from the family. None of the mothers of girls with LQTS mentioned concern about the possibility of childbirth provoking a cardiac incident.

Stage 5. The Reemergence of Uncertainty

The final stage which emerges from this study is the reemergence of uncertainty. Farnsworth, Fosyth, Haglund, and Ackerman (2006) address this issue when they state that the majority of parents in their study contended with an ongoing sense of uncertainty. The present study does not refute these findings. What is not reported in the Farnsworth, Fosyth, Haglund, and Ackerman (2006) study is the progression from the mothers' initial reaction to the diagnosis, evolving emotions and cognitions, and the progression of coping processes. All 10 mothers studied had a strong emotional reaction upon hearing the diagnosis. The mothers then developed various coping behaviors and strategies. Over time, the mothers appeared to decrease their anxiety and distress and reintegrate into their cultural milieu as a whole. However, in the back of their mind was a subtle, but not always conscious, perpetual anxiety. The anxiety appeared to increase as their children grew older and/or the mothers began to discuss their children's future upon leaving home as young adults or older adolescents. It is at this point that there was a reemergence of anxiety. This appeared to be due to the mothers' inability to monitor, restrict, and control their children's environments once they leave the home.

Finally, a few of the mothers expressed fear about the next generation. For example, one was concerned that her children may marry, not take their LQTS seriously, and give birth to children, without consideration of the effects of LQTS. Another also expressed some concern about her child needing to make decisions when he is of age to conceive the next generation.

Implications

LQTS is a life-threatening chronic disease affecting a significant portion of the population, with youth and young adults being more susceptible to its consequences. It is essential for health care providers to recognize the signs and symptoms of LQTS. Although electrophysiologists and other specialists in the field are exceptionally knowledgeable and competent, it has been revealed throughout these interviews that some health care professionals, but certainly not all, lack a clear understanding of the signs and symptoms of LQTS, even though LQTS is three times more common than childhood leukemia and occurs with more frequency than cystic fibrosis (Moss, 2009).

Ongoing education of health care workers regarding LQTS is of vital importance in order to prevent unexpected and premature death in the population.

In addition, it appears that some general practitioners or family physicians may not be fully aware of the signs and symptoms of LQTS. It is of utmost importance that family practitioners become aware of various medications and other environmental triggers that may cause a cardiac incident. For example, some over-the-counter medicines or commonly used antibiotics can have a deadly effect if prescribed without the full understanding of LQTS. In fact, this lack of understanding from the general health care community left some of mothers feeling isolated and solely responsible for the management of their children’s chronic illness (Farnsworth et al., 2006).

The mothers of these children are also presented with diagnostic and medical information which initially, and even over time, is confusing and frightening. The mothers worried about making appropriate choices for various treatments for their children. Others worried about the side effects of the medication as the children become older. Some mothers believed that the health care system is not fully on their side. These factors can leave these mothers bewildered, confused, and overwhelmed at the prospect of their child dying suddenly, even if appropriate treatment is provided. Therefore, it would be of benefit to provide parents with detailed and outlined information (Farnsworth et al., 2006) about the illness, with ongoing communication between the parents and health care staff (Dodgson et al., 2000).

The mothers interviewed in this study all appeared to have excellent and expert medical care. But even with the best medical care, the mothers were left confronting the uncertainty and the unpredictability of the illness. LQTS is not like other diseases. There are few specific benchmarks by which to measure improvement or decompensation. This leaves mothers in a precarious position regarding medical and quality of life decisions. It is important for a strong structure of both medical and emotional support to be provided for these women and their families.

The psychological effects on mothers and families of children who have LQTS can be extremely challenging. Depending on various coping skills and problem-solving abilities, some mothers fare better than others. Although there are various resources such as websites, medical staff, and the occasional conference, most of the mothers appeared to be depending on their immediate family for support. As with the need for the health care community to increase their knowledge and understanding of LQTS, it is paramount for the mental health community, especially behavioral health psychologists and hospital social workers, to gain an understanding of the unique circumstances and dynamics of coping with such a complex chronic illness. A transitional team of health care professionals, counselors, and behavioral health psychologists could provide a more comprehensive and holistic approach to assist families with LQTS to develop a clearer understanding of the illness accompanied with teaching more adaptive coping skills and abilities. Addressing the needs of families with LQTS using the biopsychosocial model would be of great service.

Most importantly, after the mother and child are sent home from the hospital or the physician’s office with the diagnosis, the main challenges are not over. The mother, her child(ren) with LQTS, and her family must contend with developmental and social issues. It is different for a child in whom LQTS is diagnosed in adolescence than it is for a child in whom it is diagnosed at birth or when very young. The adolescent or young adult make lifestyle changes and cope with a life-threatening illness. Although it was not within the scope of this study to have examined the adolescents' ability to adjust and cope to their newly revealed disease, it has been demonstrated in this study that the dynamics between the adolescent and mother are only complicated by the illness. Mothers must attempt to ensure their children's medical compliance while assisting them to evolve through developmental stages safely.

Therefore, learning and accepting the diagnosis of LQTS is only the beginning. There is a wider spectrum of services needed for mothers of children with LQTS. Although the medical field provides ongoing physical testing, checkups, and recommendations to avoid potential death, assistance in the psychological and social milieus is still lacking. Psychologists and other mental health workers are needed to provide services to mothers and their families in assisting them with how to cope with various developmental milestones, life transitional challenges, and medical compliance and understanding.

The public at large needs to be informed and educated about LQTS. Mothers find themselves needing to educate their school administrators, school nurses, teachers, and school counselors about LQTS. Some schools have automatic external defibrillators, while other schools may not recognize the need. Gyms, airports, and other public facilities would do well to outfit themselves with accessible automatic external defibrillators. Finally, some mothers and families struggled with the social stigma of having a child with LQTS. Whether these fears are substantiated or not, it is still a concern for the family. Each family has their own particular social circle and environment. Educating mental health workers, support groups, and the general public about LQTS would likely alleviate some of the unnecessary anxiety and fear that the public may have regarding long QT syndrome. With proper public education, not only will this assist mothers, their children, and their families in coping with this life-threatening illness, but it could possibly save lives by enlightening others about the signs and symptoms of and treatments for LQTS.

Limitations

The sample for this study was small and homogeneous. Although diversity of race, ethnicity, and religious backgrounds were welcomed, only 10 mothers who were White, middle to upper middle income, educated, and living in suburban or rural areas volunteered. Therefore, generalization to a larger population was compromised; it is clear that the participants do not represent a cross-section of the population at large. The mothers asked to participate had access to some of the best health care in the country; therefore, income, resources, and proximity to services were additional factors which need to be taken into consideration. Thus, demographic characteristics of the participants may have influenced the outcome of this study, resulting in an unintended selection bias.

In addition, the sample population participated on a voluntary basis. A self-selecting process was of concern. For example, less effective problem solvers and those who may not be able to cope may have opted out of the interview. There was also concern about stimulating unpleasant feelings during the interview. This was especially true for those who have an avoidant style of problem solving. However, the researcher made every possible effort to make the interview convenient and comfortable for the potential participants to avoid this concern. In addition, resources were provided and available for any foreseeable problems that may have arisen.

Due to the limited scope of this study, it is unclear as to whether children with chronic illnesses such as LQTS are more rebellious in their adolescence or whether it is simply related to related and/or parenting styles. Another factor to consider in further studies is if there is a correlation between the age at which a child is diagnosed with LQTS and the child's adjustment to the illness.

Conclusion

LQTS affected each mother in a multidimensional manner, including the biological, psychological, and social. Common psychosocial patterns emerged from these ten interviews. Mothers were impacted significantly from the news of the diagnosis. Initial reactions included attempts to assimilate large quantities of biological information, in addition to dealing with psychological reactions of disbelief and denial. Evolving emotions included feelings of fear, anxiety, sadness, loss, and guilt. Through these various stages, mothers appeared to speak honestly and wholeheartedly as to how they attempted to cope with this chronic condition on a daily basis.

It also became clear that being diagnosed with LQTS was not a one-time experience. Mothers must cope on a daily basis with the concept that their child could die suddenly. Even though their children are being treated medically and the odds of a sudden cardiac incident are reduced significantly, mothers still have to contend with a perpetual mindfulness that various triggers in the environment may generate a serious cardiac incident.

In addition to coping with the biological and psychological aspects of LQTS, the mothers must also contend with their social-cultural environments. Some mothers adjusted quite readily, while others were concerned about social stigma and their child being singled out or ostracized as vulnerable or different. A few mothers also needed to contend with their extended and immediate families' difficulty in accepting the diagnosis. Several of the mothers also described the need for greater public awareness of LQTS.

Developmental issues were also raised during the interviews. Mothers whose adolescent children recently learned they had LQTS described their children as having greater difficulty adjusting to long QT. Mothers who had younger children claimed to have less difficulty monitoring their children's medical compliance and felt that as their children grow older, they will have less difficulty coping with the illness.

Finally, after the mothers had gone through a series of stages from experiencing the initial shock of the diagnosis to evolving cognitions and emotions and coping with and integrating the concept of their children's LQTS within their particular and immediate environment, they were confronted with their children's future. Each mother expressed concern about their children gaining independence as a young adult. When discussing the future of their children's young adulthood and adulthood, a sense of uncertainty and concern reemerged. Each mother expressed concerned that she would not have the ability to monitor and oversee, on a daily basis, her child's environment related to their medical regimen. Therefore, these mothers have an additional task when it comes to allowing their children to leave the nest. They must trust and hope that they somehow instilled a sense of self-responsibility and self-care in their children, which will lead to a safer and healthier adulthood.

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APPENDIX A

APPENDIX B

APPENDIX C

APPENDIX D

APPENDIX E

APPENDIX F

APPENDIX A

Version: April 7, 2008 (vetterv-08-005813)

INFORMATIONAL FLYER/BROCHURE

A STUDY OF HOW MOTHERS COPE WITH THEIR CHILDREN'S LONG QT SYNDROME

What is the goal of this research study?

Little is known about how mothers' lives are affected after their child or children are diagnosed with Long QT Syndrome (LQTS). For example, how do mothers and families cope with their child's chronic condition and how does it affect their quality of life. Hopefully with your help, we will be able to identify the psychological and social needs of parents and families with LQTS to assist in the future development of LQTS programs.

Why are you inviting me to be a part of the study?

You are being invited to take part in this study because your child is a patient at Children's Hospital of Pennsylvania and because your child is between the ages of 6 months and 21 years of age who has been diagnosed with LQTS.

What will we be asked to do?

This study involves participating in a one time interview which will take about one to one and a half hour. The interview will be audio recorded and will consist of several questions to learn how you coped with your child's LQTS and how it has affected you and your family's lives. While the entire interview will be audio-taped, no names will be associated with the tapes. When you are finished with the interview, you will be asked to fill out a short questionnaire.

What are the benefits of taking part in this research study?

We hope that what we learn in this study may help us identify the psychological and social needs of all children and families with LQTS and benefit families in the future. Sometimes people feel good about sharing their personal information to help others.

Who do I contact if I want to help! Who do I call?

Dr. Victoria Vetter, M.D. is the principle investigator of the study. Dr. Vetter is Chief of the Division of Cardiology at Children's Hospital of Philadelphia. You may contact her with any questions you have about the study or to participate in the study at: [redacted] or E-mail her at [redacted]. You may also the contact the research coordinator, Twain Gonzales, M.Ed. M.S., at [redacted] or [redacted].

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IRB Expiration: U/A
Effective Date: 9/30/08

APPENDIX B

INVITATION LETTER TO MOTHERS

January 5, 2008

Dear Mother:

I am writing to tell you about a research project called “A Study of How Mothers Cope with Their Children’s Long QT Syndrome”. This study is hoping to identify the emotional and social needs of mothers and families who have children with Long QT Syndrome. The project is being conducted by a doctoral student from Philadelphia College of Osteopathic Medicine (PCOM) in cooperation with Children’s Hospital of Philadelphia (CHOP).

Fifteen to twenty mothers are being invited to participate in the study. We are contacting mothers who have children between the ages of 6 months and 17 years of age who are diagnosed with Long QT Syndrome. You are being contacted because your child/children have been identified as having LQTS and therefore you will be able to take part in the study. Mothers who choose to take part in the study will be interviewed by PCOM’s doctoral student and research coordinator, Twain Gonzales, M.Ed., M.S. Your interview will consist of several questions to learn how you coped with your child/children’s LQTS and how it has effected you and your family’s life. The questions are “open-ended,” which means you will be asked to give more than a “yes” or “no” answer. When the interview is finished, you will be asked to fill out a brief questionnaire which will take about 10-20 minutes to complete. While the entire interview will be audio-taped, no names will be associated with the tapes.

You may not gain any direct benefit from being in this study. However, the final results may help us to identify the psychological and social needs of mothers and their families with LQTS and benefit families in the future.

If you want to learn more about this project, please return the response card included in this packet. If you reply “yes” someone from the study will call you to describe the study in detail. Any questions you may have will be answered at this time. If you note a ‘no” reply on your response card, you will not be contacted again. If we do not get your response card within 14 days we will telephone you to see if you would like to take part in the study.

If you prefer, you can also respond by sending an email to the research coordinator at [redacted] or leave a voice mail at [redacted].

We hope that you are interested and will participate in this important study. You may call Mr. Gonzales at any time for more information at [redacted] or you may also call Dr. Victoria Vetter at [redacted]. Thank you for your time and interest.

Sincerely,

Victoria L. Vetter, M.D.

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Effective Date: 9/30/08

APPENDIX C

DEMOGRAPHIC QUESTIONNAIRE

A STUDY OF HOW MOTHERS COPE WITH THEIR CHILDREN'S LONG QT SYNDROME

Please answer the following questions or place a check mark next to the correct answer.

PERSONAL INFORMATION REGARDING YOU:

Your age ______ Gender _____ Male _____Female

Ethnic Background:

____Asian ____African American ____Caucasian ____ Indian ____ Latino(a) ___ Other ______

Estimated Combined Annual Income:

____$1.00-25,000 ____$25,000-$50,000

____$50,000-75,000 ____$75,000 and above

Level of Education:

Highest grade of schooling completed _____

Some college or trade school completed _____

Associate's Degree _____

Bachelor's Degree_____

Master's Degree _____

Doctoral Degree _____

Marital Status:

_____Married ____Single ____Separated ____Divorced ____Long Term Partnership

Are you working? ____ Yes ____ If so, how many hours? _____ No ______

Do you carry the diagnosis of LQTS? Yes_____ No _____

Does your child/children's father carry the diagnosis of LQTS? Yes_____ No _____

REGARDING YOUR CHILD/CHILDREN WITH LONG QT SYNDROME (LQTS):

How many children do you have who have been diagnosed with LQTS? _____

How many of your children who have LQTS are between 6 months and 17 years of age? _____

At what age was your child/children first diagnosed with LQTS? __________

At what age did you child/children begin treatment for LQTS? ___________

Does your child/children exhibit any symptoms of LQTS (i.e., fainting, shortness of breath, cardiac arrest…). Please list…

____

____

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

____________ Briefly describe the symptoms: ______________________

____

In what setting did this episode occur?

☐ school ☐ home ☐ sports ☐ other, please specify____________

How old was your child/children during his/her first LQT event or symptom? _______

When was your child/children's most recent event and who was present?

Date _____ (month)______(year)

Who was present?)____________________ Briefly describe the symptoms ___________

____

How many events in total (FAINTING, ARREST) has your child/children had?

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

On average, how many LQT events (frequency) has your child/children had?

☐ never ☐ 1-3 times ☐ weekly ☐ monthly ☐yearly

Has your child/children witnessed an LQT event (fainting, sudden cardiac arrest) of another family member?

☐ yes ☐ no

Does your child/children have a pacemaker or implanted cardioverter defibrillator (ICD)?

☐ Pacemaker ☐ ICD ☐ Both ☐ Neither

If you child/children has a pacemaker of implanted cardioverter defibrillator (ICD), does it create any difficult issues or problems for you? Please briefly explain________________

____

____

Is your child/ children on medication for LQTS? Yes_____ No _____ Does monitoring the medication create any difficulty for you or your child/children, if so, please explain? _________

____

____

____

Does your child/children have any other diagnosed medical or mental health problem?

☐ no ☐ yes if so:

What is the condition? ______________________________________

When was it diagnosed? ______________________________________

Any undiagnosed problems?______________________________________

Medications (current) ____

____

How do you feel about the genetic aspects of the condition?____

____

____

Has genetic testing been done? Yes _____ No ______ If so, who else has LQTS? _______________

____

APPENDIX D

The Children's Hospital of Philadelphia

CONSENT TO PARTICIPATE IN A RESEARCH STUDY

A STUDY OF HOW MOTHERS COPE WITH THEIR CHILDREN'S LONG QT SYNDROME

Study Key Name: ***HOW MOTHERS COPE WITH THEIR CHILDREN'S LQTS***

Principal Investigator: Victoria Vetter, MD Telephone:[redacted] Address: S 34th St & Civic Center Blvd. Philadelphia, PA 19104

Co-Investigator: Twain Gonzales, M.Ed., M.S. **Telephone:**[redacted]

Invitation to Participate

You, and your child, may be eligible to take part in a research study. This form gives you important information about the study. It describes the purpose of the study, and the risks and possible benefits of participating. If there is anything in this form you or your child do not understand, please ask questions. Please take your time. You or your child do not have to take part in this study if you do not want to. If you take part, you can leave at any time.

Parents or legal guardians, who are giving permission for a child, please note: in the sections that follow the word 'you' refers to 'your child' where applicable.

1. Why are you being asked to take part in this study?

You are being asked to take part in a research study on how you as a mother learned to cope with your child or children's Long QT Syndrome (LQTS). You are being invited to take part in this study because you have a child or children between the ages of 6 months and 17 years old with LQTS.

2. What is the purpose of this research study?

We are doing this study because little is known about how mothers of children with LQTS and their families cope with the LQTS. The purpose of this study is to help us know the emotional and social needs that mothers and their families may have.

This study involves one meeting to participate in a confidential and private interview. The interview will consist of questions in order to learn how LQTS has affected you and your family's lives. The interview will require approximately 60-90 minutes of your time. We expect that 15-20 mothers of children with LQTS will take part in this study.

3. What is involved in the research study?

If you agree to take part in this study, you can expect the following things to happen:

- Your child's medical history and records will be reviewed.
- A one time, 60-90 minute long interview will take place. You will be asked several questions about how you learned to cope with your child's LQTS and how you and your family's life was/is affected.
- The questions are "open-ended", which means you will be asked to provide more than a "yes" or "no" answer.
- Your answers may be as long or short as you want. Sometimes the interviewer may also ask you some short follow-up questions to be certain that what you are saying is clear.
- When the interview is done, you will be asked to fill out a brief personal history questionnaire. This will take about 10-20 minutes to complete.
- Your interview will be audio taped, but will not include your name or any other identifying information. No one will know your real identity.

4. What are the risks and discomforts/constraints of taking part in this research study?

Taking part in a research study may involve risks and discomforts/constraints or "side effects." Discomforts may include mental or emotional discomfort. This is likely not to happen. If you do feel uncomfortable, it will likely be because you will be talking about how your life has been affected by your child's LQTS.

You may also feel emotionally uncomfortable because some of the questions may be about your personal life and/or your quality of life. Although unlikely, some people may get upset in response to some of these questions. You do not have to finish the interview if it is causing you discomfort or you begin to feel too sad. If you get upset, you can simply end the interview. Ending the interview will not affect your child's or family's health care.

If you are upset, the interviewer and your child's doctor will help you find a professional with whom you can talk, or you can use the resource list which will be given to you.

5. Are there any benefits from taking part in this study?

There are no direct benefits to you for participating in this study. However, we hope that what we learn in this study may help us identify the psychological and social needs of mothers and families with LQTS and benefit other families in the future. Sometimes people feel good about sharing their personal information to help others.

6. What happens if you decide not to take part in this study?

If you decide not to participate or to withdraw from the study there will not be any penalty or loss of benefits to which you are otherwise entitled. Yours and your children's current and future medical care at Children's Hospital will not be affected if you decide not take part in this study.

7. Do you need to give your consent in order to participate?

Yes. Agreement to participate must be obtained in order to take part in this research study. There are no consequences for deciding not to sign this consent form.

8. Will confidential information be collected as part of this study?

We need to collect health information about you and your child in order to conduct this study. This includes information about you from medical records and from the procedures, interviews and tests that are part of this research. Your medical records will be reviewed to obtain information about you and/or your child's LQTS history. We will do our best to keep your personal information private and confidential. However, we cannot guarantee absolute confidentiality. Your personal information may be disclosed if required by law.

The results of this study may be shown at meetings or published in journals to inform other doctors and health professionals. We will keep you and your child's identities private in any publication or presentation about the study.

People and organizations that may inspect and/or copy your research records to conduct this research, assure the quality of the data and to analyze the data include:

- Members of the research team at CHOP;
- Medical staff who are directly or indirectly involved in your care related to this research;
- People who oversee or evaluate research and care activities at CHOP;
- People from agencies and organizations that perform independent accreditation and/or oversight of research; such as the Department of Health and Human Services, Office for Human Research Protections.
- researchers at the Philadelphia College of Osteopathic Medicine who are working with us on this study;

By law, CHOP is required to protect your health information. The research staff will only allow access to your health information to the groups listed above. By signing this document, you are authorizing CHOP to use and/or release your health information for this research. Some of the organizations listed above may not be required to protect your information under Federal privacy laws. If permitted by law, they may be allowed to share it with others without your permission.

The information collected about you or your child as part of this study will be retained for at least seven years or until the study is completed, whichever is longer. At that time, the research information will either be destroyed or all the information that identifies you will be removed from the study results and the key destroyed. All tape recorded interviews will also be erased and/or shredded. Any information collected as part of the study and put into your medical record will be kept permanently.

Use of your information in future studies

Your information may be useful for other studies on LQTS. We can only use your information again if a special committee in the Hospital, the Institutional Review Board, decides it is ok. This

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committee may want us to talk to you again before we do another study using your information. But the committee may also let us do research without talking to you again if we keep your health information private. You may also tell us that you do not want us to use your information in future studies.

9. What if you want to leave the study after you begin?

Taking part in a research study involves time and responsibilities. You need to follow the study doctor's instructions. Please consider the study time commitments and responsibilities as a research subject when making your decision about participating in this study.

You may change your mind and take back your authorization to use and disclose your health information at any time. Even if you take back your authorization, we may still use and disclose the health information we have already obtained about you as necessary to maintain the integrity or reliability of the current research. To take back your authorization, you must send a letter to Dr. Vetter. In the letter, you must say that you changed your mind and do not want us to collect any more health information about you. If you ask that we no longer collect your health information you will have to leave the study.

10. Will there by any costs to you?

There are no direct costs to you or your insurance.

11. Will you be paid for taking part in this study?

You will receive 2 movie tickets/vouchers for participating in the study as a thank you.

12. Who is funding this research study?

Yes. It is funded by the Center for Chronic Diseases of Aging at Philadelphia College of Osteopathic Medicine.

13. What if you have questions about the study?

If you have questions about the study, call Twain Gonzales, M.Ed., M.S. at [redacted] or call the study doctor, Dr. Victoria Vetter, M. D. at [redacted].

This study has been approved by a special group at Children's Hospital called the Institutional Review Board (IRB). The IRB looks at research studies like these and makes sure your rights and welfare are protected. You can talk to a person in this group if you have questions about your rights as someone taking part in a research study. You can call the Institutional Review Board (IRB) office at 215-590-2830 if you have questions or complaints about the study.

You will get a signed copy of this consent form after you sign it. You may also ask to see a copy of the full study plan (protocol).

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September 26, 2008 (vetterv08-005813)

Consent to Take Part in this Research Study

The research study and consent form have been explained to you by:

Person Obtaining Consent

Signature of Person Obtaining Consent

Date:

By signing this form, you are indicating that you have had your questions answered, you agree to take part in this research study and you are legally authorized to consent to your child's participation. You are also agreeing to let CHOP use and share your health information as explained above. If you don't agree to our collecting, using and sharing your health information, you cannot participate in this study. **NOTE:** *A foster parent is not legally authorized to consent for a foster child's participation.*

Name of Subject

Signature of Subject (if applicable)

Date

Name of Authorized Representative
(if different than subject)

Relation to subject:
☐ Parent ☐ Legal Guardian

Signature of Authorized Representative

Date

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September 26, 2008 (vetterv08-005813)

Child Assent

I have explained this study and the procedures involved to ________________ in terms he/she could understand and that he/she freely assented to take part in this study.

Person Obtaining Assent

Signature of Person Obtaining Assent Date

This study has been explained to me and I agree to take part.

Signature of Subject
(optional)

Date

For children unable to assent:

I certify that ________________ was not capable of understanding the procedures involved in the study sufficiently to assent to study participation.

Person Responsible for Obtaining Assent

Signature of Person Responsible Date

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APPENDIX E

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APPENDIX E

LQTS RESOURCE LIST

CHOP Services

Hope Lives Here.

Connelly Resources for Families (CHOP)

- (215) 590-4968
- Resources for medical information and psychological aspects of cardiac conditions
- Family Library
    - (215) 590-5778
    - The Family Library, also located in the Connelly Resource Center for Families, a quiet comfortable place where families can find healthcare, parenting and recreational information in a variety of formats.
- Family Learning Center
    - (215) 590-3661
    - Family Learning Center is a place where families can learn both the basic and specialized skills they need to help them care for their child at home

CHOP Psychiatry Department

- (215) 590-1000
- 3440 Market Street, Philadelphia PA 19104
- Behavioral Health Center

OTHER SERVICE PROVIDERS

Center for Brief Therapy – PCOM

- (215) 871-6487
- 4190 City Avenue, Philadelphia PA
- $60 for first session
- $25 for each additional

- sliding scale available
- Assessment and Treatment for Adolescents

Temple Health System – Department of Psychiatry and Behavior Science

- (215) 707-3364 (contact Deborah)
- 1316 W. Ontario Street, Philadelphia PA
- Jones Hall, 7th and 8th Floors

Counsel for Relationships

- (215) 382-6680 ext 3134
- Specialization in the following areas:
  - ☐ Family Therapy
  - ☐ Parenting
  - ☐ Children and Teens
  - ☐ Children with Special Needs
  - ☐ Anger Management
  - ☐ Diversity Issues
  - ☐ Depression and Anxiety
  - ☐ Post-Traumatic Stress Disorder
  - ☐ Grief
  - ☐ Chronic Illness
  - ☐ Psychiatry

CARE Foundation - http://www.longqt.org

- Provides online information and resources for families
- Support groups offered

SADS Foundation - http://www.sads.org/

- Provides excellent information and resources for families
- Provides networking information to connect with other families
- Offers support services

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LQT Yahoo User Group - http://health.groups.yahoo.com/group/ihfgroups/

- Provides individuals with interaction with other people with similar medical disorders.

  http://www.long-qt-syndrome.com/lqts_links.html

- message board and support groups

  http://www.qtsyndrome.ch/cgi-bin/discus/discus.pl

- message board
- **Depression Online Resources**
  - http://www.depression.com/
  - http://www.aabt.org/mentalhealth/factSheets/?fa=factSheets
  - http://www.nlm.nih.gov/medlineplus/depression.html
- **Anxiety Online Resources**
  - http://www.adaa.org/
  - http://www.aabt.org/mentalhealth/factSheets/?fa=factSheets
  - http://www.nlm.nih.gov/medlineplus/anxiety.html
  - http://www.medicinenet.com/anxiety/article.htm

APPENDIX F

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INTERVIEW QUESTIONS

1. How did you first become aware of LQTS in your family?
2. What was your reaction upon hearing the diagnosis and what did you do?
3. What has been the most difficult aspect of the diagnosis of LQTS to accept?
4. How have you worked to accept this?
5. How has LQTS affected you and your family's life?
6. How did you first adjust and cope with the illness and what would you say was the most effective means you relied on to cope?
7. Have your thoughts and feelings (and methods of coping) about LQTS and your child/children changed over time? If so, how?
8. Tell me your thoughts about your child/children's future.
9. Can you tell me about any parenting challenges you encountered with your child/children?
10. Relating to those challenges and LQTS, how did you deal with them?
11. What resources did you find most helpful in dealing with LQTS?
12. Do you feel you understand the diagnosis of LQTS? What has been most helpful in your understanding?
13. Is there anything I left out or that you would like me to know that I didn't cover?

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FOLLOW-UP QUESTIONS

14. How has the rest of your family responded to the diagnosis? Grandparents? Aunts? Uncles? Supportive or denial? If you know the "side" of the family that carries the gene, have those family members been screened? Do you feel responsible for them?
15. What are the stresses related to repeated physician appointments and testing? How could this be less stressful?
16. How has LQTS affected your child? In family? Outside family in school, activities, etc?
17. Has your child had any restrictions?
18. What are your specific concerns about your child?
    a. Sleep overs?
    b. Gym class?
    c. Class trips?
    d. Driving?
    e. College?
        i. Drinking
        ii. Staying up late
    f. Sports?
    g. Other?
19. How do you participate in child's care?
    a. Medications giving?
    b. Restrictions?
        i. Caffeine? Chocolate?

    ii. Sports?

20. How has LQTS affected your child? In family? Outside family in school, activities, etc?

21. Has your child had any restrictions?