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# Fathers of Children with Long QT Syndrome: Their Concerns and Ways of Coping

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Philadelphia College of Osteopathic Medicine

Department of Psychology

FATHERS OF CHILDREN WITH LONG QT SYNDROME: THEIR CONCERNS  
AND WAYS OF COPING

Jodi Yarnell

Submitted in Partial Fulfillment of the Requirements for the Degree of

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DEPARTMENT OF PSYCHOLOGY

**Dissertation Approval**

This is to certify that the thesis presented to us by Jodi Yarnell on the 4th day of April, 2016, 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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## **Abstract**

Congenital long QT syndrome (LQTS) is a genetic disorder of the heart that may result in syncope, seizures, and sometimes sudden cardiac death. The wide-ranging clinical presentation of LQTS can lead to fear and uncertainty in families of children diagnosed with LQTS, while treatment and lifestyle modifications can significantly reduce the level of risk. Despite the far-reaching implications of this syndrome on the lives of children and families, very little is known about the psychosocial aspects of having a child with this syndrome. The purpose of this qualitative study was to describe the experiences of fathers of children with LQTS and to investigate their concerns and ways of coping. Fathers' responses to children's LQTS have not been previously examined separate from the responses of mothers. A total of 13 fathers of children with LQTS participated in this study, six of whom took part in a one-to-one semi-structured interview and seven of whom completed an online questionnaire. Fathers experienced fear and confusion when their children were initially diagnosed with LQTS. Initial fears were moderated over time with increasing knowledge about LQTS, children's positive response to treatment and management, and fathers' perception of LQTS as a manageable condition. Background worries, however, remained related to the uncertain threat of LQTS-related symptoms developing in their children. The level of psychosocial stress that fathers experience over time may vary according to a number of LQTS-related and psychosocial variables, and fathers appeared to utilize a range of strategies to manage stress associated with their children's LQTS.

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## **Chapter 1**

### **Introduction**

#### **Statement of the Problem**

Congenital long QT syndrome (LQTS) is an abnormality of cardiac rhythm characterized by delayed ventricular repolarization and observed as a prolonged QT interval on electrocardiogram. A potentially life-threatening disorder, inherited LQTS is a heterogeneous syndrome that may result in spontaneous syncope, seizures, and sometimes sudden cardiac death (Goldenberg, Zareba, & Moss, 2008). Cardiac events associated with LQTS tend to occur more frequently in childhood, with as many as 53% of children affected with LQTS having had their first cardiac event by the age of 15 years (Zareba et al., 2003). Children and young adults who have not been diagnosed may be particularly at risk for cardiac arrest and sudden death (Hendriks et al., 2005). Previously, diagnosis of LQTS was based on electrocardiographic evaluation, while the current diagnosis for LQTS involves several tests to establish its presence, including DNA testing. Although genetic testing provides confirmation of a diagnosis of LQTS, it cannot predict the clinical course of the syndrome or the risk to the individual of future cardiac events, including sudden death (Farnsworth, Fosyth, Haglund, & Ackerman, 2006). Medical treatments and lifestyle changes can help reduce the risk of LQTS leading to potentially life-threatening events; however,

some individuals continue to experience cardiac events despite treatment and restrictions on daily life activities.

The diagnosis of LQTS can be overwhelming for all family members, who struggle with fears and uncertainty regarding the child's well-being (Farnsworth et al., 2006). Parents of children diagnosed with LQTS have many challenges to face, including an array of decisions regarding treatment options and lifestyle modifications for their children. While the uncertain and wide-ranging course and prognosis of this syndrome is likely to have a significant impact upon parents' emotional well-being, little is known about the psychosocial aspects of this condition for individuals with LQTS and their families. To better understand the experiences of parents confronted with their children's diagnosis of LQTS, emerging studies have examined the impact of this condition upon families. The psychological effects for parents whose children underwent genetic testing for LQTS were studied, and high levels of distress were reported among parents who were informed that their children were found to be carriers of LQTS (Hendriks et al., 2005). These parents reported more distress at the time of their children's diagnosis than parents of healthy children, and they remained vulnerable to higher levels of distress over time. Many parents of children with LQTS experience persistent worries and fear regarding their children's well-being and feel the need to maintain a constant watch over their children to avert catastrophe (Farnsworth et al., 2006).

While the studies examining psychosocial aspects of LQTS are limited, even less understood are the experiences of fathers and the ways in which fathers cope with and manage their children's diagnosis of LQTS. As is widespread in child

psychology research, pediatric psychology research has primarily focused on mothers, leaving the experiences of fathers less examined (Phares, Lopez, Fields, Kamboukos, & Duhig, 2005). A review of research on issues related to pediatric psychology revealed that fathers are underrepresented, even more so than when compared to research in clinical child psychology. A very small percentage of the research related to pediatric psychology included fathers only, and fewer than half of the studies reviewed included both parents in their research design. Even when both mothers and fathers were included in the research, less attention was given to the ways in which mothers and fathers may experience and cope with stress differently, without separate analyses of maternal and paternal effects (Clark & Miles, 1999).

Despite the overreliance on the mother's perspective in pediatric psychology research, this trend is changing, with increased interest in the unique experiences of fathers. More attention is also being given to the similarities and differences between maternal and paternal roles and reactions. When fathers have been included in research related to pediatric psychology, both similarities and differences emerge between maternal and paternal responses to childhood illness. In regard to affective responses of mothers and fathers in relation to childhood illness, the research is conflicting as to whether fathers experience more or less distress than that experienced by mothers when their children are ill. Several studies portray mothers as bearing the greater burden of caretaking and, as a result, experiencing stress levels higher than those experienced by fathers. Other findings, however, suggest that both mothers and fathers experience heightened levels of distress, while their particular concerns and sources of stress may vary. Additionally, factors influencing parental

adjustment to their children's illness, such as social support, coping strategies, and family cohesion, have been shown to vary for mothers and fathers (Dewey & Crawford, 2007).

The considerable variability in the ways that mothers and fathers adjust to their children's health condition may be understood within the context of the disability-stress coping model (Wallander & Varni, 1998). This model was developed to explain the experiences of families of children with chronic illnesses in terms of stressors, risk factors, and resistance factors. Research in pediatric psychology has demonstrated the potential for chronic health conditions in children to cause distress in families, including the presence of increased depression, anxiety, and posttraumatic stress disorder (Gudmundsdottir, Gudmundsdottir, & Elklit, 2006; Streisand et al., 2008). This model is based on previous theories of adjustment to chronic disease and specifies the risk factors that may increase vulnerability to stress and the resilience factors providing a protective function. Intrapersonal factors, such as cognitive appraisals of family members, have been incorporated into many conceptual models of coping, as have socioecological factors, such as social support and family cohesion (Gudmundsdottir et al., 2006). Fathers' ways of coping and adaptation examined within the broader context of a conceptual model provides a framework for understanding particular strengths and vulnerabilities that may be targeted in future interventions.

### **Purpose of the Study**

The purpose of this study is to examine fathers' experiences and reactions to their children's LQTS and to investigate the coping strategies used to manage those

reactions. The study will explore fathers' reactions, concerns, and perceptions associated with children's LQTS, as well as the different styles of coping that fathers engage in to manage stress. The potential findings of this study could have implications for the development of targeted interventions to help fathers of children with LQTS. The psychosocial functioning of fathers is likely to be important for their children's physical and emotional adjustment and likely to play a role in the way children adapt to their illness (Ryan, Ramsey, Fedele, Wagner, Mullins, & Chaney, 2011). These findings may also contribute to a broader understanding of the experiences of fathers of children with serious health conditions within the context of a risk-resistance coping model.

## **Literature Review**

### **Introduction**

As previously mentioned, congenital long QT syndrome (LQTS) is a genetic disorder that may increase a person's risk of sudden, uncontrollable, and life-threatening arrhythmias, which, in some cases, can lead to sudden cardiac death (Delaney, Mittal, & Sherrid, 2009). It is characterized by a family of genetic mutations known as channelopathies because the affected genes encode directly and indirectly for the ion channels of the surface of each heart muscle cell. In LQTS, this malfunction of cardiac ion channels can affect and disrupt the electrical activity of the heart, characterized by a prolongation of the QT interval. A long QT interval, while of no consequence in and of itself, places individuals at risk of developing torsade de pointes, a dangerous form of arrhythmia. In the United States, the incidence of congenital LQTS is estimated to be 1 in 2,500 to 5,000 (Delaney et al., 2009). The

most common presentation of LQTS is syncope, while in 12% of symptomatic patients, aborted cardiac arrest or sudden cardiac death is the first manifestation of the disease. Syncope, although nonfatal, is associated with comorbidities, such as trauma, and is a much more frequent cardiac event than aborted cardiac arrest or sudden cardiac death (Liu et al., 2011). Recurrent syncope is a powerful predictor of subsequent fatal or near-fatal events, independent of the corrected QT Interval (QTc) (Liu et al., 2011). While syncope is the most common symptom of LQTS, approximately 6 to 13% of affected individuals undergo cardiac arrest or sudden cardiac death before the age of 40 years when untreated (Ackerman, 1998). Congenital LQTS is one of the leading causes of sudden cardiac death in children, who are more at risk than adults of developing symptoms. LQTS is believed to be the cause of sudden unexplained infant death in about 5 to 10% of cases.

### **Diagnosis of LQTS**

Diagnosis of LQTS is challenging and ultimately made based on the medical history of the child, electrocardiographic evaluation, and subsequent DNA testing (Hendriks et al., 2005). Patients can be referred for an evaluation for many reasons, and individuals may present with a range of symptoms, including syncope, seizures, and aborted sudden cardiac death, while others remain asymptomatic. Male individuals with the disease often present during preadolescence, while female individuals present later (Locati et al., 1998). Asymptomatic patients may be referred for an evaluation as a result of the identification of the disease in a first-degree relative. For both symptomatic and asymptomatic patients, the QT interval can be normal on the initial presentation, making diagnosis even more difficult (Delaney et

al., 2009). Asymptomatic individuals can be particularly difficult to diagnose, as their QT interval is more likely to have a normal duration as compared to that of symptomatic patients. A scoring system is used to aid in diagnosis and takes into account various factors, such as gender and prolongation of the QT interval; however, this approach may result in missed diagnoses for borderline scores (Delaney et al., 2009).

### **Treatment**

The risk of cardiac arrest and sudden death associated with LQTS can be reduced with proper treatment after diagnosis (Hendriks et al., 2005). Once a diagnosis is made, the treatment of LQTS involves consideration of the patient's presentation of symptoms and may involve lifestyle changes, beta-blocker medication, or the placement of an implantable cardioverter-defibrillator (ICD). Patients who have already had aborted cardiac arrest are considered to be at the highest risk for a future occurrence, and medical treatment and ICD placement are recommended (Moss et al., 2000). For patients without prior cardiac events, treatment involves beta-blocker medication and lifestyle modifications. Beta-blockers are associated with a pronounced reduction in the risk of a first cardiac event among children and adolescents with LQTS (Goldenberg et al., 2008). Therapy with beta-blockers has been demonstrated to be quite effective, but it is genotype-dependent and can result in unwanted side effects, such as mood swings, depression, and fatigue (Farnsworth et al., 2006). Beta-blocker therapy may have important limitations among patients with LQT2 and LQT3 and among patients who experience syncope during beta-blocker therapy. Lifestyle modifications include avoiding

known triggers of cardiac events that prolong the QT interval and, depending on the type of LQTS, may include the avoidance of swimming, exercise, stress, and auditory stimuli. The management of young athletes and physically active individuals creates dilemmas for children and their families given the higher risk of potentially life-threatening cardiac events associated with exercise in certain genotypes (Kapetanopoulos, Kluger, Maron, & Thompson, 2006). Not all types of LQTS have known external or internal triggers, making decisions regarding lifestyle choices difficult.

Treatment involves decision-making guidelines based on a series of risk stratification strategies that take in such variables as QT duration, gender, and type of LQTS. These risk stratification strategies, however, are limited, and the treatments themselves involve their own risks. The most common treatment for LQTS, beta-blockers, can have side effects. While beta-blockers are often effective, syncope, aborted cardiac arrest, and LQTS-related death continue to occur while patients are on prescribed beta-blockers, particularly in those who were symptomatic before starting this therapy (Moss et al., 2000). While ICD placement is recommended for the patients with LQTS at highest risk for cardiac events, risks are also involved in its use, and risk-benefit judgments therefore must be considered (Sherrid & Daubert, 2008). Disagreement within the medical community regarding recommended treatments is likely to further complicate the ability of family members to feel secure in knowing which approach to take. In addition to the fear and anxiety that can result from the diagnosis alone, the complex decisions regarding treatment of LQTS are likely to lead to feelings of confusion and uncertainty in parents. In particular, fathers

who may have less involvement with their children's healthcare providers may feel uncertain in making appropriate decisions regarding treatment. Fathers may feel unprepared for making decisions without sufficient knowledge regarding their children's condition and prognosis.

### **Psychosocial Aspects of LQTS**

Not surprisingly, the diagnosis of LQTS in children is associated with high levels of distress in parents whose children have LQTS; however, few studies have examined the psychosocial aspects of families living with LQTS. When a child is diagnosed with LQTS, parents are confronted with the reality of a potentially life-threatening cardiac abnormality in which some children may remain asymptomatic while other children may die suddenly. With a broad and uncertain prognosis, parents reasonably experience fear and doubt regarding their children's well-being.

Hendriks et al. (2005) examined parental adjustment to the diagnosis of LQTS by assessing the short-term and long-term psychological effects on parents whose children underwent predictive genetic testing. Of parents who were informed that their children were carriers of LQTS, 50% showed clinically relevant distress levels shortly after the diagnosis was given. Follow-up findings revealed that 18 months after the time of initial diagnostic confirmation of LQTS, levels of distress remained high, with one third of parents continuing to demonstrate clinically high levels of disease-related anxiety and one quarter demonstrating high levels of depression. These findings regarding longer term adjustment were less favorable than those of other studies of parents whose children had undergone predictive genetic testing (Broadstock, Michie, & Marteau, 2000). The higher level of distress

displayed by these parents may be explained by the difficulty in adjusting to their children's vulnerability to fatal arrhythmic events (Hendriks et al., 2005).

**Sources of distress.** The sources of distress that parents of children diagnosed with LQTS experienced were examined further (Hendriks et al., 2005). Parents reported several areas likely to be contributing to elevated levels of distress. Parents reported their greatest fear as the persistent threat of symptoms of LQTS suddenly occurring in their children, and they maintained ongoing vigilance for these symptoms (Hendriks et al., 2005). Most of the children were receiving treatment, and while almost two thirds of the parents felt they had thoroughly considered treatment decisions, most parents continued to feel uncertain regarding the efficacy of the treatments selected.

Another source of anxiety for parents was concern regarding the future of their children (Hendriks et al., 2005). The diagnosis of LQTS may place limitations on a child's activities that impact their future career and relationships, as well as may result in stigmatization. More than half of the parents worried about the approach of puberty and anticipated diminishing control over their adolescents' actions and medical adherence. In addition to these sources of worry and concern, parents reported dissatisfaction with the support and the information about the disease they received and were more likely to have disease-related and general anxiety higher than that experienced by other parents. Parents reported feeling more knowledgeable and informed about the nature of the disease than their physicians. Farnsworth et al. (2006) examined perceptions and reactions of parents towards LQTS, given the far-reaching clinical spectrum and resulting ambiguity regarding prognosis and

treatment. Parents' concerns fell into four main areas: fears of death, quality of life, uncertainty, and implications for practice. Mothers' and fathers' concerns were not reported separately in order to determine possible differences in their particular areas of concern. Parents expressed fears regarding the prognosis of their children's condition and a persistent sense of concern regarding their children's perceived precarious state.

**Variables related to high levels of distress.** Hendriks et al. (2005) examined variables related to high levels of distress in parents of children diagnosed with LQTS. Along with dissatisfaction they had with the information provided by physicians, parents with low levels of education were more likely than other parents to experience higher levels of disease-related anxiety. Higher disease-related anxiety was also associated with parents who had longer familiarity with the disease and had experienced sudden death in their family. Longer familiarity with the disease and dissatisfaction with given information were also associated with high levels of general anxiety and depression. These findings were confirmed in a qualitative study of mothers of children with LQTS (Gonzales, 2009). Mothers who had knowledge of LQTS before it was diagnosed in their children, who had experienced their own cardiac event, and who had reported a difficult medical experience upon diagnosis were more likely than other mothers to have higher levels of distress.

**Quality-of-life issues.** Quality of life for families with children with LQTS is primarily impacted by the lifestyle choices made in order to avoid triggers that are most likely to put their children at risk. Parents expressed a desire to make balanced, well-informed decisions regarding lifestyle choices for their children. Parents appear

aware of the impact of their decision-making behavior on their children's adjustment, wishing to reduce children's worries by not being excessively cautious themselves. Lifestyle modifications can be particularly difficult when a diagnosis is made in adolescence, and families have to contend with the prospect of restricting their teenagers in ways that impact not only their daily activities but also their career goals. Loss and sadness may be felt by parents who grieve over the quality of life that they perceive their child has been denied (Gonzales, 2009).

Quality of life is also impacted by treatment decisions, and families face complex choices with various implications. Parents expressed concerns about the long-term side effects of medications, as well as about the observed changes in their children's mood and behaviors. ICD therapy was also a difficult decision for parents who contemplated its long-term impact on their children's lives, as well as potential risks and device complications. When children approach adolescence, parents may struggle most significantly with medical adherence issues and experience their children's outright refusal to take medication (Gonzales, 2009). Although parents worry about a reduced quality of life associated with children's LQTS, some parents reported an improved quality of life, associated with changes in their attitudes towards life accompanied by a reevaluation of priorities. This phenomena is consistent with attitude changes reported by people who survive life-threatening experiences (Arteaga & Windle, 1995).

**Implications.** The need for knowledgeable healthcare providers who can assist families in the management of LQTS is underscored by the complex and difficult decisions that families face. Parents expressed the need for ongoing support

in managing their children's condition and in assessing responses to treatment, such as intolerance to beta-blockers, and treatment of illness. Parents want more research and information on different aspects of LQTS treatment, such as the long-term impact of beta-blockers and ICDs on children as they grow, and more studies of families as they adjust to their children's diagnosis.

**A comparison of psychosocial effects in families with LQTS and asthma.** Only one study examined the psychosocial aspects of LQTS as compared to another chronic health condition, in this case asthma (Giuffre, Gupta, Crawford, & Leung, 2008). Like families with children with LQTS, families of children with asthma often report medical fears and must monitor environmental triggers to avoid symptom exacerbation. Specifically, a comparison of children with asthma and children with LQTS in terms of their anxiety and medical fears was conducted. Mothers of children were also included in the study, and a parent questionnaire was administered to assess parental levels of anxiety. Results indicated that children with asthma had significantly higher scores on medical fears and fears of death, while children with LQTS may have been more likely to underreport their anxiety. Children with LQTS, who were more likely than children with asthma to display internalizing problems, may have more difficulty in openly discussing their fears and worries (Giuffre et al., 2008). Mothers of children with LQTS were found to report significantly higher levels of state anxiety compared to mothers of children with moderate and severe cases of asthma, a finding consistent with other studies of families with inherited LQTS.

## **Uncertainty and Its Psychological Effects**

Studies specifically examining the psychosocial aspects of LQTS suggest that the uncertainty associated with a child's diagnosis is likely to lead to increased levels of distress and anxiety in parents and has been identified as a major stressor affecting families of children with chronic conditions (Dodgson et al., 2000). The inability of parents of children with LQTS to predict outcomes with certainty leads parents to worry about their children and the possibility of the development of potentially fatal symptoms. Furthermore, parents' skepticism regarding the effectiveness of treatment decisions despite having thoroughly considered various options may contribute further to their stress as they face uncertain outcomes (Hendriks et al., 2005). Even when treated with medication or ICD placement, children may still experience symptoms. The less satisfied parents feel with the knowledge they receive regarding their child's diagnosis and treatment, the more uncertain parents are regarding their child's symptoms and prognosis.

While parents are significantly affected by the uncertain outcomes involved in their children's diagnosis of LQTS, most research related to the concept of uncertainty has focused on adults with illness. Relatively few studies have investigated the influence of uncertainty in childhood chronic conditions (Dodgson et al., 2000). Uncertainty is described as a state in which a person cannot adequately define or categorize an event because of a lack of sufficient cues or information and is viewed as a major component of one's illness experience (McCormick, 2002; Mishel, 1988). According to Mishel (1981, 1990), uncertainty is a multidimensional concept that is present throughout diagnosis, treatment, and prognosis of illness and is

separate from its psychosocial outcomes. These complex events associated with illness often include ambiguous, vague, unpredictable, unfamiliar, inconsistent, and unknown factors that underlie one's experience of uncertainty (McCormick, 2002). Probability is an attribute shared by these factors and can impact the degree of uncertainty perceived in a given situation (McCormick, 2002). For example, certain situations may involve known strategies to avoid negative outcomes, while the actions taken do not guarantee that negative outcomes will not occur. Uncertain situations such as these, which involve an unclear probability, are likely to be experienced by parents of children with LQTS who may take extra safeguards to protect their children without a guarantee of their safety.

High levels of uncertainty are associated with increased anxiety and depression and impact one's psychosocial adaptation and family functioning (Dodgson et al., 2000; McCormick, 2002). Studies that have examined uncertainty in families revealed that both mothers and fathers whose children experienced intermittently unpredictable symptoms experienced more distress than mothers and fathers of children whose condition was more predictable (Dodgson et al., 2000). Parents of children with epilepsy, who are often faced with a constant feeling of uncertainty, reported ongoing anxiety related to each seizure experienced by their children, including fear of their children dying during the seizure (Mu, Wong, Chang, & Kwan, 2001).

Symptoms of depression may also be experienced by parents who are faced with the unpredictable and ambiguous nature of their children's condition and the potential impact it has on family functioning and sources of social support (Mu, Kuo,

& Chang, 2005). As mentioned previously, 25% of parents of children who were found to be carriers of the LQTS mutation had high levels of depression 18 months after the test outcome (Hendriks et al., 2005). A survey of 50 parents of children with epilepsy reported a sense of depression, anger, guilt, frustration, and hopelessness (Thomas & Bindu, 1999). Prevalence data among parents of children with a heterogeneous sample of childhood disorders suggest that while most parents do not meet criteria for clinical levels of depression, nearly half of parents report significant elevations in depressive symptoms and may be at risk (Silver, Westbrook, & Stein, 1998).

To examine fathers' uncertainty and consequential coping strategies, a group of 31 fathers of children undergoing cancer treatment completed Mishel's Parent Perception of Uncertainty Scale (PPUS) and the Jalowiec Coping Scale (JCS; Sterken, 1996). Results indicated that the father's age, the child's age, and the length of time since diagnosis all correlated with paternal uncertainty, with younger fathers whose children were also younger being most likely to experience information received as vague and ill defined. Not surprisingly, fathers were also more likely to experience uncertainty when their children had been diagnosed for only a short period of time. A significant relationship was demonstrated between uncertainty and coping. Fathers who did not use a confrontive coping style were more likely than fathers who did use a confrontive coping style to experience uncertainty and a lack of clarity regarding the information received about their children's treatment and the system of care. Given the medical complexity of LQTS and its treatment, this finding suggests that healthcare providers need to continuously assess fathers' understanding,

as they may not verbalize their perception of information received as vague or ambiguous (Gonzales, 2009).

The relationship between paternal uncertainty, coping patterns, and depression was examined in fathers of children with epilepsy (Mu, 2005). Results indicated that paternal uncertainty was positively correlated with depression. Coping behaviors of maintaining family integrity, keeping an optimistic outlook, preserving social support and self-esteem, and understanding the medical condition were associated with lower levels of uncertainty and were important for fathers' adaptation. Paternal coping patterns of preserving social support and self-esteem and understanding the medical condition were primary predictors of fathers' depression, suggesting that facilitation of effective coping strategies by fathers may help reduce uncertainty and improve family adaptation.

### **Impact of Parents' Coping on Children's Adjustment**

The psychosocial adjustment of mothers and fathers of children with LQTS and other childhood conditions not only is important for their own well-being, but also has an influence on children's coping and adaptation. Family factors may be more important in determining children's psychosocial adaptation than disease characteristics or severity (Bender et al., 2000). Factors associated with the psychological adaptation in children with mild to moderate asthma were examined and included measures of the psychological adaptation of the family and disease-related variables (Bender et al., 2000). Results indicated that the emotional climate of the family was the strongest predictor of the child's psychological adaptation, while disease-related factors were not.

Factors related to parental adjustment should include a consideration of the experiences of both mothers and fathers of children with LQTS who are likely to each play a role in making decisions and managing their children's health. While studies on parental experiences of childhood illness have traditionally focused on the perspectives and responses of mothers, research indicates that fathers play an important role in setting the emotional climate in the home and that children may be particularly sensitive to fathers' levels of distress (Eiser, 1990; Robinson, Gerhardt, Vannatta, & Noll, 2007). Higher levels of parental stress and perceived child vulnerability among fathers of children with chronic illnesses significantly predicted youth uncertainty (Ryan et al., 2011). As fathers' experiences and influence are better understood, healthcare providers will be better equipped to address fathers' particular needs and facilitate positive adjustment among family members.

### **Fathers and Chronic Childhood Illness**

**Fathers' role and influence.** Relatively less is known about fathers of children with chronic health conditions as compared to the experiences of mothers. A number of societal and theoretical developments over the past century have contributed to a significant increase in the attention given to fathers and their inclusion in both developmental and pediatric psychology research (MacDonald & Hastings, 2008). Women's career roles outside of the home have contributed to a shift in fathers' involvement at home, with both parents assuming a more equally divided role in care-taking responsibilities (MacDonald & Hastings, 2010). Of fathers with children under the age of 5 years, 20% were the primary caretaker in 2010, and the number of married fathers who stay at home to take care of their children has been

increasing (U.S. Census Bureau, 2013). Theoretical developments have placed a greater emphasis on the role of the father in infant attachment and as part of a larger system of family members (MacDonald & Hasting, 2008; Rutter, 1972; Seligman & Darling, 2007).

Increased awareness of the role of fathers in their children's lives has led to greater inclusion of fathers in research and examination of their influence on child developmental processes (Lewis & Lamb, 2003). Research on paternal involvement indicates that children with highly involved fathers demonstrate higher cognitive skills, demonstrate greater empathy towards others, and display a greater internal locus of control as compared to children whose fathers are less involved (Lamb, 2010; Pleck, 1997; Radin, 1982, 1994). Furthermore, stronger connections between fathers and their children are likely to have a positive and significant influence on the quality of the relationship and the well-being of the child (Brotherson, Yamamoto, & Acock, 2003). Research suggests that the relationship that fathers develop with their children may be jeopardized when the family is confronted with the diagnosis of a chronic illness in a child (Clark & Miles, 1999). The emotional response that fathers experience associated with their children's health condition may influence the quality of their interactions and their level of involvement with their children. Fathers may fear becoming too attached to their infants diagnosed with a life-threatening condition, limiting their level of involvement (Clark & Miles, 1999). Youth sports is one area in which fathers often are highly involved in their children's lives, but children's participation in sports may be restricted by such health conditions as LQTS (Coakley, 2006). Children considered particularly at risk because of previous

LQTS-related cardiac events may be excluded altogether from competitive sports (Kapetanopoulos et al., 2006). Fathers, who often monitor their children's involvement in sports activities, may be affected by this aspect of their children's development (Coakley, 2006).

Pediatric psychology research investigating fathers and children with chronic health conditions lags behind developmental and clinical child research in including fathers in research designs (Lamb, 2010; Phares et al., 2005). Researchers have called for giving greater attention to the concerns of fathers of children with special healthcare needs, analyzing separately for maternal and paternal effects when fathers are included, and including fathers in family-based interventions (May, 1996; Phares et al., 2005). While studies specifically examining LQTS from the father's perspective do not exist, an emerging body of research about fathers of children with chronic health conditions is beginning to provide a greater understanding of their individual experiences (McNeill, 2004).

**Fathers' responses to childhood health conditions.** The parenting concerns and coping strategies of fathers of children with chronic health conditions were compared with the concerns and coping strategies of fathers of well children using the Hymovich Family Perception Inventory (Hovey, 2003). Children with chronic health conditions included in the study had cancer, cystic fibrosis, or juvenile rheumatoid arthritis (JRA). While both groups of fathers reported similar types of concerns, the degrees of concern experienced by fathers of children with chronic health conditions were significantly greater. Fathers of children with chronic health conditions worry more about their children's health and are more concerned about being adequate

caregivers and providers. Similar to previous studies, these fathers were most concerned about their ability to recognize important changes in the health condition of family members (Cayse, 1994; Hovey, 2003). Financial issues were also indicated, with fathers expressing concerns about the cost of healthcare and maintaining insurance coverage. Fathers of children with chronic health conditions indicated the importance of communication and involvement with family members. They were more concerned regarding the perceived lack of control they have over circumstances affecting their families. A comparison of the clinical and nonclinical groups of fathers indicated that fathers of children with chronic health conditions are significantly more likely to report crying as a strategy for coping, in contrast to other findings (Cayse, 1994). This finding suggests that although fathers may be reluctant to openly reveal their emotions to others, some fathers express their distress in a more private manner. The fathers of children with chronic conditions were just as likely as the fathers of children who were well to use problem-solving coping strategies. Fathers may experience financial stress caused by various factors, including reduced income as a result of missed days of work and extra costs involved in their children's care. The degree to which financial issues are a concern may vary, however, in relationship to family earnings.

Interesting differences emerged in the concerns of fathers of children with chronic conditions in respect to family income (Hovey, 2006). A higher percentage of fathers with an annual income of less than \$50,000 were concerned about their ability to care for their family and obtain necessary information. These fathers expressed greater financial stress regarding health insurance costs along with general

money concerns. In terms of coping strategies, fathers in the higher income group were more likely than the fathers in the lower income group to change their expectations regarding their children and family life as a coping strategy. The higher income fathers also found this strategy more helpful than did other fathers. Although both groups of fathers used prayer as a coping strategy, the lower income fathers were much more likely to find it helpful. Whether or not family income is a factor affecting families of children with LQTS is not clear; however, this specific concern has not been reported by mothers and fathers in previous studies. Fathers with higher paying jobs may have greater flexibility, allowing them increased opportunities to participate in their children's clinic visits and receive information directly from healthcare providers.

**Fathers' conflicted reactions.** Clark and Miles (1999) explored the experiences of fathers of infants newly diagnosed with complex congenital heart disease. Six of the fathers of children with congenital heart disease were Caucasian, one was Asian, and one was African American. Semi-structured interviews were conducted to explore fathers' experiences with a focus on the time of the child's initial diagnosis and treatment. The findings demonstrate the conflicting nature of their reactions. At the time of the child's diagnosis, these fathers experienced intense sadness and a sense of loss over the normal child they had hoped to have. Along with their sadness, many fathers also reported feelings of joy that came along with the birth of their children and becoming a father. Similar to findings from other studies, these fathers experienced tension regarding their internal distress and their reluctance to outwardly reveal these emotions in order to be a source of strength for their

families. Most fathers felt threatened by their growing attachment to their child. Conflicting emotions were also revealed between the fathers' need to maintain control and their sense of powerlessness. To cope, fathers attempted to maintain a semblance of normalcy in their daily routines and work.

**Fathers' coping and positive reappraisal.** A holistic picture of fathers' experiences was revealed following interviews of 22 fathers of children with JRA (McNeill, 2004). Semi-structured interviews were conducted to explore fathers' experiences of caring for a child with a chronic health condition. Among those interviewed, three of the fathers temporarily stayed at home as the primary caregiver when their children were newly diagnosed. The study included stepfathers and single, separated, divorced, and remarried fathers. Of the fathers who refused participation in the study, most were visibly minorities or of lower socioeconomic status and had declined to participate because they were too busy or uninterested. Consistent with results of other studies, fathers were profoundly affected by their children's illness, while a sudden onset of symptoms, uncertainty regarding the diagnosis, and risk to survival contributed to more intense and, at times, catastrophic emotions. Fathers had difficulty dealing with the uncertainty resulting from the unpredictable course of JRA, struggled to make meaning of events, and felt the need to remain in a perpetual state of readiness. Periods of increased stress and uncertainty affected fathers' levels of work productivity. Similar to other groups of fathers, they had difficulty openly expressing the depth of their emotions to others, leading some fathers to feel isolated. Fathers often assumed a protective and supportive role, reluctant to appear vulnerable or to burden their wives with their own

worries. With limited outlets to express their emotions, they relied mainly on self-support through prayer, exercise, distraction, and cognitive strategies.

Fathers coped with their children's illness by actively maintaining an optimistic attitude (McNeill, 2004). Searching for positive meanings helped fathers to make sense of their children's illness, identifying the positive aspects arising from an otherwise difficult reality. Fathers regarded themselves as taking a practical and pragmatic approach to dealing with their children's illness, which they viewed as balancing out their partners' more emotional responses. Furthermore, fathers viewed themselves as trying to balance their efforts to make life as normal as possible for their children, despite their protective worries. Consistent with other findings, these fathers experienced closer relationships with their children and families as they redefined their life priorities (Neil-Urban & Jones, 2002).

The fathers in McNeill's (2004) study, depending on the severity of their children's condition, were often confronted with their children's pain and suffering. Fathers reported that managing their children's pain was one of the greatest challenges that they experienced. As mentioned previously, children with LQTS may be either asymptomatic or otherwise healthy with proper medical treatment and spared from the physical pain that is more characteristic of other childhood disorders such as cancer and JRA. The uncertainty inherent in the unpredictable nature of JRA is a feature it holds in common with LQTS, and parents are likely to experience the anxious anticipation of suddenly developing symptoms. As for parents of children with LQTS, the uncertainty inherent in their

children's condition resulted in the fathers' perceived need to be on constant guard for recurring symptoms.

### **Conceptual Framework**

The disability-stress coping model was developed to provide a conceptual framework for understanding the experiences of families and children with chronic illnesses by identifying factors that influence their adjustment (Wallander & Varni, 1998). It is based on previous theories of adjustment to chronic disease, family coping, and cognitive appraisal and coping and is intended to be applicable to a wide range of pediatric chronic disorders (Gudmundsdottir et al., 2006). The model takes into account the interaction between various disease, intrapersonal, and environmental factors that influence the manner in which parents cope and adapt to the stressors brought about by a child's illness (Katz, 2002a). The disability-stress coping model identifies both risk and resistance factors that either increase risk for adjustment problems or serve as protective factors that minimize adjustment difficulties. Investigation of these factors may help explain the individual differences that are found among parents of children with chronic illnesses (Wallander & Varni, 1998). Risk and resistance factors are viewed as modifiable and potential targets of empirically supported interventions aimed at parents of children with chronic conditions.

Wallander and Varni (1998) identified two categories of risk factors that may increase vulnerability to parental distress: the disease parameters/functional care strain and psychosocial stress. Resistance factors that may buffer the effects of childhood illness include intrapersonal factors, such as self-esteem and self-efficacy;

socioecological factors; and stress-processing factors, such as cognitive appraisal of events.

**Risk factors.** Parental adjustment to children's LQTS may be influenced by the presence of risk factors which increase vulnerability to parental distress such as the clinical severity of their children's condition and associated stressors.

***Disease parameters and functional care strain.*** Research studies have examined the degree to which disease variables, such as diagnosis, symptom severity, visibility of disease, and degree of impairment, impact parental adjustment. As previously mentioned, LQTS is characterized by a wide spectrum of clinical severity that depends on the individual's age and gender and on the genetic mutation that is causing the long QT. Family members who share the same mutation will not necessarily experience related symptom severity (Kimbrough et al., 2001). The principal events associated with LQTS are syncope, seizures, ventricular tachycardia, and, less commonly, cardiac arrest or sudden cardiac death. Nature of onset and course and risk of fatality are also important factors in understanding family functioning and parental adjustment. The onset and course of LQTS can vary widely among children, and risk of fatality continues to be refined through identification of risk factors and more precise stratification of patients (Berul, 2008). Families who experience a sudden onset of symptoms in their children and uncertainty about the diagnosis may experience greater distress (McNeill, 2004).

***Psychosocial stress.*** Psychosocial stress is another variable that can increase risk of parental adjustment difficulties. Events that produce increased stress can be directly or indirectly related to the child's condition and can include events requiring

hospitalization or limitations placed upon the child's current or future functioning. Whether or not the parents of children with LQTS who display more cardiac symptoms experience higher levels of distress than the parents whose children display fewer symptoms has not been empirically studied, although a substantial number of parents showed signs of anxiety even when their children never experienced any symptoms (Hendriks et al., 2005). Risk stratification is used to guide treatment of patients but cannot predict with certainty the course of symptomatology for an individual patient (Nemec, Hejlik, Shen, & Ackerman, 2003). This uncertainty may result in worry in many parents who fear symptoms developing in their children. Fathers may experience more uncertainty at the time of their children's diagnosis, when there may be more confusion and limited knowledge about their children's condition and its implications (McNeill, 2004; Sterken, 1996).

The management of LQTS can be a source of stress for parents who must make complex decisions about their children's treatment and lifestyle. Parents strive to reach a delicate balance between protecting their children from events that may place them at risk and not diminishing their quality of life. Fathers may be particularly concerned with providing their children with as normal a life as possible (McNeill, 2004). A fatherly tendency to encourage an attitude of confidence and curiosity in his children may be strained by a new set of precautions. Worries about his child's future in regards to career, dating or marriage prospects, and stigmatization are another source of stress (Hendriks et al., 2005). Limitations on certain types of activities may interfere with children's career goals (Farnsworth et al., 2006).

Illness in a child with LQTS can be another cause of concern for parents (Farnsworth et al., 2006). Dehydration and fever secondary to illness in a child can increase risk for cardiac events. Extra care needs to be taken to avoid medications that can further prolong the QT interval. Parents also worry about their children's anticipated adherence to medical treatment regimens as they approach adolescence (Hendriks et al., 2005). As mentioned previously, while a greater number of mothers than fathers, in general, tend to report family life stress, fathers who do experience high family stress may be more prone to adjustment difficulties. Of the few studies that have examined parents' responses to their children's LQTS, none have explored the particular sources of stress most concerning to fathers.

**Resistance factors.** Resistance factors, such as parents' cognitive appraisals and coping efforts in response to children's LQTS, may influence positive adjustment in parents.

**Stress processing.** The disability-stress coping model draws upon a cognitive-phenomenological theory of psychological stress and coping in which stress is viewed as existing not within an external event, but within a transaction between a person and his or her environment (Folkman, Lazarus, Dunkel-Schetter, DeLongis, & Gruen, 1986; Lyon, 2012). Two concepts central to stress theory are cognitive appraisal and coping, which are described as critical mediators within the person-environment transaction (Folkman et al., 1986). Individuals appraise events as either threatening with the potential of loss or challenging with the possibility of benefit or mastery. Coping efforts involve both emotion-focused and problem-focused coping styles and are influenced by the individual's appraisal of a particular

situation. According to the uncertainty in illness theory, uncertain situations are evaluated by the appraisal categories of danger and opportunity, and coping efforts are activated to mediate the relationship between appraisal and adjustment (Mishel, Padilla, Grant, & Sorenson, 1991). Mishel et al. (1991) conceptualized that the nature of the appraisal will influence the type of coping strategy activated, such that threatening situations may call for emotion-focused strategies, while problem-focused coping strategies are activated when a situation is viewed as an opportunity that can be altered. However, the relationship between appraisal categories and coping efforts may vary according to the particular situation and the related variables, such as the severity of the individual's health condition.

A number of studies have examined mothers' and fathers' styles of coping in response to their children's health condition (Dewey & Crawford, 2007; Katz, 2002; Mastroyannopoulou, Stallard, Lewis, & Lenton 1997). Less attention has been paid to the specific appraisals in parents of children with a chronic illness, which are thought to influence selected coping strategies (Wallander & Varni, 1998). Further investigation regarding illness-specific appraisals in both mothers and fathers may increase understanding of the psychological processes involved in parental adjustment and gender differences that may exist in the ways parents process stressful events.

The extent to which fathers of children with LQTS view their children's situation as either threatening or challenging may influence their coping behaviors and adaptation. Uncertainty results from being unable to categorize and determine the meaning of an event because of a lack of a predictable pattern of symptoms and

outcome (Mishel, 1988). Events that are viewed as uncertain are evaluated as dangerous until the threat of potential harm is minimized or mastered through the implementation of coping strategies (Mishel, 1988; Sterken, 1996). A confrontive coping style was associated with less uncertainty among fathers of children diagnosed with cancer, whereas fathers who used a less aggressive approach to manage their situation were dissatisfied with the information they received about their children's condition and prognosis.

***Intrapersonal factors.*** Intrapersonal factors, such as self-efficacy, optimism, and problem-solving ability, may account for variations in maternal and paternal adjustment to stress (Wallander & Varni, 1998). Parents who perceive themselves as competent in managing their children's illness are less likely to experience adjustment difficulties (Noojin & Wallander, 1997). Although intrapersonal factors are likely to contribute to parents' overall adjustment, research investigating the intrapersonal factors that influence paternal adjustment is limited. Fathers may be more likely than mothers to display confidence regarding their ability to cope effectively, particularly during periods of illness (Mastroyannopoulou et al., 1997). Despite their perceived competence in coping effectively during periods of worsening symptoms, fathers were less likely to view themselves as competent in the medical management of their children's illness (Knafl & Zoeller, 2000). No current research specifically examines intrapersonal factors associated with fathers' adjustment to LQTS, while exploration in this area could provide information about fathers' sense of optimism, self-efficacy, and problem-solving regarding the management of their children's LQTS.

***Socioecological factors.*** Socioecological factors refer to the social environment and include such variables as family support, marital satisfaction, and family conflict (Wallander & Varni, 1998). Research suggests that mothers may be more likely than fathers to experience increased dissatisfaction with their relationship with their spouse, and low family cohesion is associated with mental-health adjustment difficulties for both parents (Dewey & Crawford, 2007; Mastroyannopoulou et al., 1997). Having a shared view in which parents share perspectives on the management of their child's illness is considered a family strength that may lead to better adjustment (Knafl & Zoeller, 2000). Fathers may be more likely than mothers to adopt a minimizing perspective regarding their children's illness in which the negative impact of their children's condition is downplayed (Knafl & Zoeller, 2000). Parents of children with LQTS face a number of important decisions regarding their children's medical treatment and lifestyle management, and may benefit from adopting a shared perspective regarding the management of their children's health condition. Differing spousal perspectives could potentially lead to increased conflict, while interventions aimed at enhancing mutual support may lead to improved outcomes (Knafl & Zoeller, 2000).

### **Summary**

LQTS is a potentially life-threatening arrhythmia syndrome that may result in spontaneous syncope, seizures, and sometimes sudden cardiac death, especially in undiagnosed children and young adults. Studies examining the psychosocial adjustment of parents of children with LQTS are limited in number but suggest that many parents experience fear and uncertainty associated with a continuous threat of

developing symptoms in their children. The need to address not only the physical management but also the psychosocial management of childhood health conditions has been established in the literature while the psychosocial implications of living with LQTS continue to be understood. Furthermore, the need for an increased family orientation in pediatric research has led to greater inclusion of fathers in studies involving children's serious health conditions. The purpose of this study was to investigate psychosocial aspects affecting families living with LQTS by describing the experiences of fathers of children who have been diagnosed with this condition. Identifying the nature of fathers' concerns as described from their unique perspectives may promote broader understanding of psychological effects of LQTS and support a family-centered and holistic approach to treatment.

### **Glossary**

QT - The QT of LQTS refers to an interval between two points (Q and T) on the common electrocardiogram (ECG, EKG) used to record the electrical activity of the heart.

QTc - The corrected QT interval (QTc) estimates the QT interval at a heart rate of 60 beats per minute. The QTc allows comparison of QT values over time at different heart rates and improves detection of patients at increased risk of arrhythmias.

Pediatric psychology - The study of the interrelationship between the psychological and physical well-being of children, adolescents, and families.

Ventricular arrhythmia - An abnormal, usually rapid heart rhythm that is caused by abnormal electrical impulses in the lower chambers of the heart, called the ventricles, and is often life threatening.

## **Chapter 2**

### **Research and Interview Questions**

#### **Research Questions**

1. What are the personal experiences of fathers related to having a child with LQTS?
2. What are the particular concerns, challenges, and coping strategies of fathers of children with LQTS?

#### **Interview Questions**

1. How did you first become aware of LQTS in your family?
2. Tell me about your initial reaction when your child was diagnosed with LQTS.

What

did you do? What events led up to your child's diagnosis?

3. What have been the most difficult aspects of the diagnosis of LQTS for you? How have you dealt with these challenges? How have you responded the same as or differently from your spouse? What have been the most difficult decisions you have faced?
4. In what ways has having a child with LQTS affected you? Your relationship with your child? Your relationship with your other children? Your relationship with your spouse?
5. Has having a child with LQTS affected the way you view your role as a father? In what way?
6. Describe your greatest concerns regarding your child and his or her condition.
7. Has LQTS restricted your child from extracurricular or physical activities? If so,

in what way? Have restrictions affected your relationship with your child? If so, how?

8. To whom do you turn for emotional support? Social support? Are there any other resources that you feel would have been/ would be helpful?
9. Describe your experience with your healthcare team from the time of diagnosis until the present.
10. What was it like for you to talk about your experience today?
11. Is there anything that I haven't asked you that you would like to comment on or share?

## **Chapter 3**

### **Methods**

#### **Overview**

This study examined the reactions, perceptions, and coping strategies of fathers of children with congenital LQTS. The presence of LQTS in a family can place the emotional well-being of children with LQTS and of their parents at risk. Research suggests that mothers and fathers differ in their perceptions of, reactions to, and coping with the illness of their children. Information regarding the experiences of fathers of children with LQTS currently is limited. A qualitative study was used to examine fathers' experiences and viewpoints in relationship to the topic of the study.

#### **Design and Design Justification**

A qualitative design was used to investigate the reactions, perceptions, and coping patterns of fathers of children with LQTS. Two methods of data collection were used in this study. The first method involved in depth interviews using a standardized semi-structured format designed to gather detailed information about the participants' individual experiences. The interviewer was a doctoral student in a clinical-psychology graduate program. The standardized open-ended interview called for a semi-structured format. Participants were asked a series of identical questions, but the questions were worded to elicit open-ended responses. Open-ended questions allowed the participants to contribute as much detailed information as they wished and also allowed the interviewer to ask probing questions as a means of follow-up. A

series of questions and probes was used to examine four main areas: initial reactions to the diagnosis; beliefs and attitudes regarding their children's condition; coping styles; and family, social, and community supports.

The second method of data collection involved a web-page-based survey using an open-ended questionnaire format that fathers completed online (Appendix A). The online questionnaire was an abbreviated version of the semi-structured, one-on-one interview and included eight questions.

### **Participants**

A total of 13 individuals participated. Six of the participants were fathers who participated in one-on-one interviews. Seven of the participants were fathers who completed an online questionnaire. This sample of fathers was selected to facilitate examination of the impact of childhood LQTS on fathers and to explore their experiences.

### **Inclusion and Exclusion Criteria**

#### **Inclusion Criteria**

The participants consisted of fathers living inside or outside of the United States who were 18 years and older with children diagnosed with LQTS syndrome for at least 1 year. The participants included biological or step-parents living in the household of the child. The children either tested positive for LQTS mutation or had a confirmed clinical diagnosis and were 1 to 18 years of age.

#### **Exclusion Criteria**

The study did not include divorced fathers or unmarried fathers who assumed only partial custody of the child.

## **Recruitment**

To recruit participants for the one-on-one interview, the child's physician mailed a letter describing the study to prospective candidates for the research study (Appendix B). A return postcard or e-mail was included to confirm interest in participating in the study. Upon confirmation of interest, the interviewer made a phone call to the prospective participant to schedule the time and date of the interview. Participants were also recruited for one-one-interviews online through announcements of the study placed on LQTS-related websites and social network sites (Appendix C).

Participants were recruited for a web-based questionnaire through announcements placed on LQTS-related websites and social network sites. A brief summary describing the purpose of the study was posted on websites and social media sites with a link to the web-based questionnaire (Appendix D). The questionnaire was created at SurveyMonkey.com.

## **Measures**

The Personal History Questionnaire was completed by fathers who participated in one-to-one interviews (Appendix E). The questionnaire included demographic information regarding age, gender, level of education, primary language, ethnicity, marital status, household income, and occupation. History of LQTS in the family and questions specific to onset and symptoms also were included. A standardized open-ended interview was used and explored four main areas: (a) initial reactions to the diagnosis, (b) beliefs and attitudes regarding their

children's condition, (c) coping styles, and (d) family, social, and community supports.

### **Procedures**

Interviews were conducted at participants' homes or by phone. The interviewer explained the purpose of the interview, addressed terms of confidentiality, and obtained informed consent. The interviewer explained the format of the interview, indicated the approximate length of the interview, and asked participants if they had any questions before beginning the interview. Each interview took 1 to 1½ hours to complete.

Each interview was recorded, transcribed verbatim, and checked for accuracy. Pseudonyms were assigned to all participants, and names of places were changed to maintain confidentiality. The data were reviewed and coded by a team of three individuals enrolled in the doctoral clinical psychology program. The coders had successfully completed a graduate-level course on qualitative research and were trained in coding using data analysis techniques as described in Corbin and Strauss (2008). Data analysis involved open coding in which the data were broken down using a labeling process for each piece of dialogue from the transcribed interviews. Links were then made among the data using axial coding, in which relationships between the codes are derived. Concepts, core categories, and themes were developed through the process of constant comparison. After all the interviews were completed, the team members were provided with written transcripts of each interview, and each transcript was independently coded by each member of the coding team to bring multiple perspectives into the analysis. The team met at regular

intervals to validate the findings by comparing themes derived from the original transcripts. At each meeting, one or two interviews were reviewed by all team members.

The online questionnaire was developed on SurveyMonkey.com. A link to the questionnaire survey was posted on various LQTS-related websites with a description describing the study. The same coding process was used to analyze the data gathered from the online survey. A copy of each survey was provided to team members to code, and then a meeting was held to review the coded surveys. The team continued to meet at regular intervals until all interviews were reviewed, taking a total of 12 hours.

## **Chapter 4**

### **Results**

Each interview was analyzed by each coder independently according to three phases: open coding, axial coding, and selective coding (Strauss & Corbin, 1990). Coding refers to the process of ascribing labels that identify, name, categorize, and describe phenomena in the text. After each coder independently coded each interview, all three coders met to compare and contrast their findings. A basic framework was used to identify relationships between core categories, essential concepts, and theoretical models to explain the findings.

#### **Participant Characteristics**

These findings were obtained from a total of 13 fathers of children with LQTS, six of whom participated in one-to-one interviews and seven of whom participated in an online survey. Demographic data obtained from fathers participating in the one-to-one interviews are described in Table 1. Because fathers participating in the online survey did not complete a demographic questionnaire, information was gathered indirectly via their online survey responses and may provide a limited representation of these participants. Of the six fathers who were interviewed in person or via telephone, four fathers had one child diagnosed with LQTS, one father had one child with a confirmed LQTS diagnosis and one child with a borderline diagnosis of LQTS, and one father had two children with LQTS. Of the seven fathers who participated in an online survey, five fathers had one child with

LQTS, one father had two children diagnosed with LQTS, and one father had three children with LQTS.

The circumstances under which fathers first became aware of LQTS in their families varied widely and are outlined in Table 2. Of the 13 fathers interviewed and surveyed, 10 fathers first learned of LQTS in their families at the time that their children were diagnosed. The remaining three fathers had prior knowledge of LQTS at the time of their children's diagnosis as a result of an earlier discovery of LQTS in another family member. Furthermore, circumstances varied in the manner by which a diagnosis of LQTS was discovered in children. For one father, LQTS was first discovered when his daughter participated in a screening at her school for student athletes, but otherwise had not experienced any cardiac symptoms. Two fathers first learned of the diagnosis from the discovery of a very low heart rate in utero prior to their children's birth. One father's daughter was diagnosed with LQTS subsequent to being bitten by a tick and experiencing unspecified cardiac symptoms. Five fathers had children diagnosed after presenting with syncopal episodes. One father discovered his daughter had LQTS after she had a cardiac arrest. Children of participants were various ages when first diagnosed with LQTS, with the youngest children being diagnosed at birth and the oldest child receiving a diagnosis at the age of 17 years. Overall, nine of the children diagnosed with LQTS were male and nine of the children with LQTS were female.

Table 1

*Summary of Participant Demographics*

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Pseudonyms	Age	Ethnicity	Educational level	Salary range
Bob	53	Caucasian	Bachelor's degree	\$75,000 and above
Gary	46	Caucasian	Some college	Unknown
Michael	57	Caucasian	High-school diploma	\$75,000 and above
Ted	59	Caucasian	Bachelor's degree	\$75,000 and above
Kevin	46	Caucasian	Bachelor's degree	\$75,000 and above
Amir	Unknown	Asian/Middle East	Master's degree	\$25,000 or under

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*Note.* Demographic information was not gathered from fathers who completed on-line questionnaires.

Table 2

*Summary of Parent and Child LQTS-related Factors*

Carrier	Gender of child(ren)	Age at time of dx	LQTS <sup>a</sup> discovered by	# of events	Type of treatment
Father	Female	3 months	EKG	0	Beta-blocker
	Male	10 years	Syncope	1	Beta-blocker
Father	Female	5 years	Tick bite	0	Beta-blocker
Mother	Female	15 years	Screening	0	Beta-blocker
Father	Female	Birth	Low heart rate	7 or more	ICD <sup>b</sup> ; Beta-blocker
Mother	Female	Birth	EKG <sup>c</sup>	0	Beta-blocker
DiNovo Mutation	Female	<1 year	Low heart rate	1 or more	ICD; Beta-blocker
Unknown	Female	Unknown	Family member	1	ICD; Beta-blocker
	Male	9 years	Seizure	1	Beta-blocker
Mother	Male	Birth	Family member	0	Unknown
Unknown	Female	Unknown	Syncope	1	ICD
Father	Male	Unknown	Syncope	3 or more	ICD
Mother	Male	8 weeks	Family member	0	Unknown
Unknown	Male	Unknown	Syncope		ICD
Unknown	Female	Unknown	Cardiac arrest	2	ICD

<sup>a</sup>Long QT syndrome.<sup>b</sup>Introcadioverter Defibrillator.<sup>c</sup>Electrocardiogram.

## **Fathers' Responses to LQTS**

### **Fear, Worry, and Confusion**

Fathers experienced a wide range of emotions in response to their children's diagnosis of LQTS. Their initial emotional reactions were generally more intense and drastic than they became over time. When first informed of their children's medical diagnosis, several fathers felt an immediate sense of fear, worry, and confusion. Bob's son was diagnosed at the age of 10 years after a fainting episode during a school assembly. He was rushed to the hospital when the nurse at the school thought he remained unconscious too long. Bob recounted hearing the news with his son at his side in a hospital room: "When we met with the cardiologist at [the hospital], sitting there with a 10-year-old who was very active and everything, and me not knowing anything about it, and she said, 'Well, here's the potential symptoms. You could have fainting spells, or you can die.'" Bob described his initial reaction of fear in response to a condition unknown to him and with a potentially life-threatening and broad clinical spectrum: "Fear of the unknown ... a lot of worrying about him and what he was going to have to deal with as he went forward."

Bob echoed the sentiments of several fathers, whose struggle to grasp the meaning of a diagnosis with such wide-ranging implications aroused fear and concern. Ted's daughter had a pacemaker implanted into her abdomen when she was 2 days old because of her very low heart rate in utero. In describing his initial reaction to his daughter's diagnosis, Ted recalled, "At first, we were pretty scared by that diagnosis ... we didn't really know what it meant, never heard of anything like this before. She was in the newborn intensive care unit for 2 weeks before we were able to

take her home, so it was a bit traumatic primarily because it was something that we never heard of.” The confusion that accompanies the LQTS diagnosis is perhaps most notable when the condition has just been discovered, as families try to quickly assimilate new information without any existing knowledge of the condition to provide a frame of reference. Fathers who want to understand the meaning of the diagnosis for their own children’s well-being are faced with reconciling the medical complexity of a syndrome with varied levels of risk. “I was lost. I’ve never heard of it,” said Gary about receiving his daughter’s diagnosis. “I was basically confused and concerned.”

Kevin’s daughter was diagnosed with LQTS when she was born and remained in the hospital for a week in the neonatal intensive care unit (NICU). He described the experience as terrifying: “One of the things that’s terrifying for both a parent and anyone that’s got long QT is that the first thing you hear is that adrenaline and alarms and sudden loud noises are bad, and the NICU is nothing but constant alarms and loud noises, and as a parent you’re just terrified that all these alarms going off are going to cause some sort of a problem, cause an adrenaline spike can kill your kid.” Kevin described the period immediately after his daughter’s birth as extremely stressful, fearing his daughter’s seemingly precarious condition while feeling completely powerless to protect her. The first couple of weeks after his daughter was brought home from the hospital were also fraught with complications that resulted in more stress. Kevin described confusion that resulted from his daughter being placed on a heart monitor that was fitted imprecisely: “It was absolute torture having our baby on that heart monitor., They put it on incorrectly and instructed us on how to put

it on incorrectly, so for a solid 2 weeks, the alarm went off every 15 minutes. It was just constant false alarms, and when you hear those things going off, and again, the last thing you want for an LQT patient is an alarm.” Kevin acknowledged that more information in the beginning would have reduced the level of confusion and anxiety that he and his wife experienced during those initial weeks: “If proper information had been available and given at the right times it would have been so much easier, um,... but you don’t know what you don’t know when you’re going through it.”

### **Shock and Disbelief**

For some fathers, the sudden knowledge that their children are at risk for life-threatening symptoms is met with physical distress, shock, or denial. One father expressed his physical upset upon the realization that his child’s recent symptoms, which had been instinctively assumed to be directly caused by the child’s own activity, were actually caused by a serious underlying condition: “I felt sick when I realized his ‘passing out’ was not from overexertion or dehydration (he was active in soccer, basketball, scouts, band) as was diagnosed by doctors but from LQTS.” The sudden realization that their active, healthy children are at risk for potentially life-threatening consequences can be shocking. For one father, this difficult task of comprehending his daughter’s diagnosis was met with doubt and denial. Michael’s daughter, Lily, was identified with LQTS after the discovery of an irregular EKG finding during a health screening for athletes at her high school. When physicians first suspected that Lily’s unusual EKG might be caused by a serious underlying condition, Michael found himself inclined to search for a more innocuous explanation. As Michael stated, “It wasn’t a clear-cut that yes, that you have it ... it

was ... that her readings were on the borderline, so we thought maybe it was ... at the time she was on some medicine ... and so I thought ...well, maybe that affected the reading.” In addition to the ambiguous diagnostic findings, Michael’s skepticism towards the physicians’ preliminary impressions was underscored by the fact that prior to the health screening at her school, his daughter had never experienced any serious health problems. He recounted his struggle to fully comprehend a medical explanation that appeared inconsistent with his reality and unlikely for a teenager who had been healthy all her life: “I thought, no that can’t be. I’m sure that she’s fine ... so I guess ... a little bit of denial ... just because we hadn’t really seen any of the symptoms when they explained it to us.” After Lily’s mother, Shawna, was tested and found to have LQTS also, confidence in the diagnosis was still reserved until confirmed by genetic testing. Michael stated, “I still thought, well, Lily has never shown, I didn’t understand it, yet what it was and how it is genetic ... but then ... when the genetic testing came back positive and then we knew what we were dealing with ... I felt nervous in that ... I knew that I was always going to have to watch for this, and I just didn’t want it to happen.”

### **Guilt**

Of the 13 fathers who were interviewed or surveyed, 10 first learned of LQTS in their families through their children’s diagnoses. Once LQTS was confirmed in their children, testing was conducted to determine if other members of the family were carriers. In several cases, neither parent had experienced any symptoms throughout their lives, and learning that they were the carrier came as a surprise. For other parents, the diagnosis explained earlier events in their lives, such as syncope,

for which they had never had a medical explanation. For some fathers, finding out that they had LQTS and that they had passed along the gene to their children brought feelings of guilt. Gary said, “It was upsetting that I was the carrier.” When fathers are not the carriers, they may find supporting their spouses to be a challenge. One father noted his greatest challenge was “dealing with my wife’s guilt about ‘giving it to our son.’” Fathers, particularly those who are not affected by LQTS, may feel like outsiders who are helpless to protect their spouses and children who are affected by LQTS. Kevin, whose wife had been diagnosed with LQTS 3 years prior to the birth of their child, is concerned that one day his daughter will resent her parents for allowing LQTS to be passed to her.

### **Disappointment**

Kevin was the only father interviewed who had a family member diagnosed with LQTS prior to his child’s birth. Different from the other fathers interviewed who had no knowledge of LQTS prior to their children’s diagnosis, Kevin prepared himself for the possibility of his daughter being born with LQTS, while he hoped that she would be unaffected by the family gene. He acknowledged that he and his wife “went in knowing that it was a possibility, hoping that it wasn’t going to be the case. It was disappointing to find out that it was, but it was something that was manageable.” Kevin’s response reflected the disappointment and sense of loss that fathers may experience when their hopeful expectations as new parents are suddenly met with a different and potentially worrisome reality.

## **Relief**

While the discovery of LQTS was an emotionally difficult experience for all fathers, the ascertainment of a medical reason for unexplained symptoms came as a relief for some. Several fathers also now had an explanation for undiagnosed symptoms of their own or of other family members. When asked to describe his immediate reaction, one father recalled a mixed response: “Both fear and relief. Fearful because my child had a heart problem. Relief because I realized almost immediately that it was a likely explanation of my syncope episodes when I was younger.” Another father recounted, “Actually a sense of relief, as we knew what had caused her cardiac arrest and that an ICD could help keep her alive.” For these fathers, the diagnosis provided an answer for a condition that could be properly treated and managed.

## **Daunting Obstacles: One Father’s Account**

Amir’s daughter, Laila, was delivered via an emergency Caesarean section when her heart rate began to drop during the last trimester of pregnancy. Only hours after she was born, Laila began to have difficulty breathing and was eventually transferred to a cardiac intensive care unit, where an abnormality was detected in her heart. After 7 weeks of testing in search of an explanation for the cardiac abnormality, physicians were still unable to diagnose Laila’s condition. Amir, who is of greater Middle Eastern descent and living in his native country, began scanning the results of the medical tests to a physician in the United Kingdom whom he was able to contact through his family and friends living there. Eventually, the abnormality of his daughter’s heart was confirmed to be caused by something wrong with the QT

interval, but still a diagnosis remained beyond reach. Ultimately, through a great deal of research and the use of Facebook, Amir came in contact with physicians in a clinic in the United States who said they would arrange for genetic testing to confirm a diagnosis of long QT if Amir could find a way to transport blood samples to the United States. Although not a simple task, samples eventually were sent, and genetic testing confirmed that Laila had LQTS. The physicians in the United States told Amir that his daughter should receive an ICD. Thus, Amir was posed with other seemingly insurmountable obstacles: the lack of a medical facility able to perform this procedure in his country along with his inability to afford the costs of travel to the United States and of the recommended treatment. When asked about his response to these events and his daughter's eventual diagnosis, Amir recounted, "So it was very, very difficult for me to comprehend even all this ... it was like a nightmare for me. I used to research about this condition all day long on the Internet in the office, then sometimes I had to go and do the night shift for my daughter cause my wife had a C-section, she was at home, she cannot go and attend the baby so I had to go to the hospital and stand all night at the door and keep looking at her vital signs and I used to cry, yes, and I used to ask God like, 'Why me?' So sometimes my belief in life, my life in goodness, kindness was lost."

## **Managing LQTS**

### **Knowledge and Understanding**

Most fathers developed a more positive view of their children's diagnosis over time as their knowledge of LQTS increased and their initial fears became less extreme. After the initial diagnosis, families took an active approach in learning as

much as they could about their children's diagnosis by educating themselves about LQTS through research on the Internet and becoming involved in social media and online LQTS groups. Four of the six fathers interviewed relied upon their children's mothers, whom they described as the primary seekers of information and as being most involved with LQTS support groups, as their source of knowledge. Mothers more often attended physicians' appointments, where medical information was shared and questions answered. Though mothers, more than fathers, were viewed as having more thorough and complete knowledge of LQTS, some fathers also sought information independently, primarily through websites, social media, or newsletters.

Although mothers are more active in collecting information on LQTS through their own self-directed research, fathers are active consumers of that information. By educating themselves, fathers feel empowered to take increasing control over the impact that LQTS has on their children and the entire family. For Kevin, knowledge helps to sort out a complex and unfamiliar medical diagnosis so that families can act in the best interests of their children, while he believes a lack of knowledge can be detrimental. He stated, "If you at least know, you can do something about it ... the worst part about it is the unknown, and if you know what you're dealing with and you know what you're supposed to do, then it becomes manageable." Kevin is an analytical thinker whose approach to solving problems is to make the best decision possible given the information that is known. When asked if he ever questions whether he is making the right decisions for his daughter, Kevin responded in straightforward terms: "Based upon all the information that we have received, what we're doing, i.e., following precautions by administering daily medications and

carrying an AED [automated external defibrillator], is the best that we can do for our child, and that just becomes part of the routine.”

Becoming better informed about the diagnosis and known treatments reduced Gary’s initial confusion and offered a direct and active approach to LQTS management. He said, “There’s a lot of things I was confused about, but now, to me, it’s pretty straightforward ... and if we don’t treat it something could happen ... if we do treat it, it prevents something from happening ... so it took the confusion away. I was basically confused and concerned, but once you start reading, you understand somewhat of what has to be done, and what doesn’t have to be done, what you watch for, what you don’t watch for.” By increasing their knowledge, fathers feel better equipped to make informed decisions about their children’s activities. Gary explained further, “I just know that there’s certain things that she can’t do ... and we just don’t do it. I just go around them. ... If she wants to ride her bike, we ride her bike. If she wants to play basketball, we play basketball. ... If the basket hoop is in the sun, we’ll play for a little bit, and then we’ll get out of the sun for a little bit, then we’ll play. ... It’s just all common sense of the knowledge of her situation.”

Similar to other fathers, Michael described how knowledge may help reduce fears as families learn how to proactively reduce risks associated with LQTS through modifications to daily routines and habits; “Yeah, it’s a watch. I feel we’re educated now and we understand and we know what to watch for, and because we’re taking the other steps and making sure [of] the simple things, like, make sure she has bananas and potassium and things like that, and the electrolytes are replenished and that she

has her beta-blocker and gets plenty of rest and avoids stressful situations, then we should be fine. So that's what we're trying to do."

Receiving a confirmed diagnosis not only allowed families to take a proactive approach to treating their children, but also opened the door to identifying other family members possibly affected by LQTS. Once their children were determined to have LQTS, families began the process of investigating their family genetic histories. Ted shared his thoughts about the positive aspects of receiving a diagnosis. "Learning about it as early as we did, it's kind of been a bit of a blessing because we're able to ... go through our whole family and ... make sure everybody got tested ... what have we gotten from it is just we've been able to share this with all family and the members of the family that need to be aware of it are aware of it and ... are taking the right precautions. It's saving lives down the line."

Amir's experience was quite different from the experiences of other fathers living within the United States because of the lack of medical knowledge and expertise in his country regarding the highly specialized area of LQTS. After his daughter's birth, Amir took matters into his own hands, extensively researching on the Internet to find out what would explain his daughter's heart problem. He said, "Over time I researched so many research papers, so many studies, so many medical conditions, that now I feel a bit accomplished that I was solely able to get my daughter diagnosed and treated, which I guess someone less literate or someone in a different profession might not be able to do because I am an IT professional. I have a good educational background, and I am also an Internet nerd. I always try to go on the Internet and find and research everything about everything." Once the diagnosis was

confirmed through genetic testing conducted in the United States, Amir experienced a sense of relief that he now understood the cause of his daughter's heart abnormality and the consequential appropriate treatments, as well as a sense of accomplishment.

### **Lifestyle Changes**

Lifestyle changes and some restrictions are important aspects of managing LQTS. Specific recommendations depend on several genetic and clinical factors and are designed to avoid situations or activities known to trigger cardiac events. Lifestyle decisions can be complicated because triggers associated most commonly with one type of LQTS may also trigger cardiac events for individuals with another type of LQTS. Exercise is a potential trigger for cardiac events in LQTS, and lifestyle considerations often involve decisions about children's participation in sports (Ackerman, 2015). The participation of athletes with LQTS in competitive and organized sports activities is guided by the medical recommendations of the 36th Bethesda Conference. While recommended guidelines aim to reduce risk to young athletes, the absolute risk to any one individual choosing to participate competitively cannot be determined with certainty and may be low for some individuals (Maron & Zipes, 2005). Outside of competitive sports, many children with LQTS wish to participate in recreational and noncompetitive sports activities. Recognizing the health benefits of exercise, parents want their children to maintain physically active lifestyles. The American Heart Association provides recommendations regarding the participation of children with cardiac conditions in noncompetitive sports activities; however, these recommendations are meant to be viewed as general guidelines rather than strict standards. While established recommendations offer practical and

reasonable guidance regarding children's participation in competitive and recreational activities, families still are faced with making the best decisions for their children.

When asked about the ways in which their children's activities have been restricted by LQTS, the responses provided by fathers were varied. Factors including significant QT prolongation and history of cardiac arrest were the most significant factors impacting the level of restrictiveness that children experienced. Ted's daughter, Tina, was diagnosed with LQTS and received a pacemaker at birth because of her long QT interval and a low heart rate. She has been restricted from competitive sports activities, according to Ted, "from Day 1," when she entered elementary school. Tina was often required to sit out from activities during her physical-education class. Ted described the social and emotional consequences of being excluded from the normal schedule of activities: "If Tina has to stand on the sidelines while all the kids are running around playing soccer or whatever, she stands out and ... in elementary school it's probably not so bad, but when you get to middle school and especially high school nobody wants to be standing out in the crowd, nobody wants to be not normal." Restrictions may be viewed as primarily imposed by outside institutions, such as schools, rather than by parents themselves. Gary stated, "I don't restrict her. I mean the school, it's the school policy with the way the state's set up, I believe, that they're not allowed to participate in the state test for gym ... She could probably do the state tests, like the presidential tests, like the mile run ... but they won't allow it, the state won't allow it or somebody won't allow it ... but I'd be willing to bet she could probably do close to a mile right now." Gary acknowledged the need for safety precautions, while also bringing attention to the

unfortunate social consequences of children being treated differently: “I understand, believe me, I understand, it’s my daughter, but at the same token they ask, ... ‘Why am I sitting here holding the stop watch and calling out times?’ ... They know... they know something is there.” Another father commented on the limitations imposed by LQTS when speaking about his three sons who are diagnosed with LQTS, all three of whom have ICDs implanted. In addition to standing out during physical-education activities, his children do not spend the night at friends’ houses. Children’s fears may also result in self-imposed restrictions, as one father spoke about his son who now worries about the consequences of overexerting himself during physical activity.

Many children are restricted from rigorous physical activity, while still able to participate in other sports of lesser intensity. One father discussed how his very active and fit 22-year-old daughter had to stop rowing and playing netball competitively after going into cardiac arrest when she was 17 years old: “She now does yoga and Pilates instead of competitive rowing and plays netball in a slow social team. But she still gets out and does her thing.” Even though Michael’s daughter, Lily, had an advantage on her basketball team because of her height, she did not continue to play after her diagnosis. She still participates on the cheerleading squad, however, one father commented on his reliance upon medical professionals in determining in which activities his children can and cannot participate: “We followed doctor advice. My son did not play flag football because of LQT. He did participate in Ultimate Frisbee. My daughter participated in both volleyball and lacrosse.”

## **Proactive Lifestyle Management**

Several fathers discussed intentionally guiding their children's activities in a desired direction. Particularly when diagnosed at an early age, parents may proactively guide their young children towards developing interests in activities that are known to carry less risk for future cardiac events. Bob discussed how his daughter, diagnosed soon after her birth, was impacted differently from his son, who was diagnosed at the age of 10 years: "I think with Kaylee it's probably restricted her more than it did our son just because being diagnosed at 3 months old, we just directed her away from things that we knew that they preferred she didn't do, like soccer, things that are real running and track. ... So it's limited her, but I don't know that she's aware that it's limited her." Whereas older children may already be actively involved in organized sports at a competitive level when a diagnosis is discovered, young children may be more amenable to their parents' efforts to shape their interests and behaviors. Kevin has known his 3-year-old has LQTS since her birth. He and his wife have given considerable thought as a couple to their approach to managing LQTS and plan to take a very deliberate approach in molding their daughter's interests and activities. Kevin explained, "Ella is going to be the best female golfer that's ever lived. She's going to be one heck of a piano player, she's going to have outlets and do things that are safe, and it's just the way it is. ... She'll ride horses, she'll play golf, she's not going to be a competitive, she'll play little kids soccer maybe, ... not sure about that yet, but she's not gonna play competitive soccer, she's not going to be a competitive swimmer." Fathers want their children to have

opportunities for meaningful engagement in a variety of activities, but always within the boundaries of safety and minimal risk.

### **Living as Normal a Life as Possible**

When discussing the potential impact that LQTS would have on their children's lives, several fathers discussed wanting their children to live a normal life, not to be treated differently, and to enjoy, as much as possible, full and active lifestyles. Bob does not want LQTS to have a significant influence on or control over the way he and his family choose to live their lives. Having been a competitive athlete through college, Bob values the role of organized sports and his children having the opportunity to participate as competitive athletes. He and his wife have been physically active and involved in sports throughout their lives, and their three children have followed in their footsteps. Bob remained supportive of his son's athletic endeavors after he was diagnosed with LQTS at the age of 10 years and did not stop him from continuing to play competitive sports. Bob has continued to live an active life himself, even after learning, subsequent to his son's diagnosis, that he has LQTS, while never having experienced any cardiac symptoms. He stated, "We've tried not to make it have any real impact on how we live our lives." When asked about the most challenging aspects of living with LQTS, Bob remarked that, fortunately, his family has not had to face potentially more difficult circumstances associated with LQTS. Other than following routine precautions, he acknowledged that he has been fortunate that his children have not been affected by cardiac events for most of their lives or required more invasive procedures. Over time, Bob and his family have become rather accustomed to the precautions embedded in the family's

daily routine: “I think the biggest thing we’ve had to do, they’ve all been active, including I’m active in sports, like for Ryan and for Kaylee, Dr. C lets them participate in certain sports, but we have to have an AED with us and we have to go to every practice, so just taking the normal precautions I guess ... I think we’ve just gotten used to doing it over 14 years of just doing it.”

**Finding a healthy balance.** The importance of remaining in good physical health is stressed by Bob, who wants to avoid unintended consequences that could result from overly limiting the activity of his children. A healthy balance is sought between living a full and active life while taking necessary precautions. Bob explained, “I’d say we’ve tried to make sure that our kids, and including myself, stay active, within reason ... from a physical perspective, I don’t want them to sit around and do nothing and then have a lot of other medical problems that would eventually go along with that. So, trying to be as normal as possible would be the best way to put it.”

**Child-centered decision making.** When families must consider whether or not to allow their children to participate in sports and other physical activities, one father says the simple path is to avoid the risks by just saying no. Michael’s daughter, Lily, is on the cheerleading team at her high school and is passionate about competitive cheerleading. The cardiologist working with Michael’s family has given his approval for Lily to continue cheering as long as she avoids situations in which her heart rate would remain elevated for a prolonged period. When Lily’s family first explained to her cheerleading coaches her condition and the precautionary measures she would have to take, there was some concern. As Michael put it, “The easiest way

is to say, no you just can't do any more sports ... just avoid it in general, but knowing how much it meant to Lily and ... how she hadn't had any symptoms or anything, the cardiologist was okay with it." Michael's family carefully considers these lifestyle decisions with respect to the importance the children attach to expressing their talents and passions through sports and team involvement.

Kevin does not want his 3-year-old daughter to be held back from enjoying the exhilaration of new learning experiences similar to those experienced by any child her age. He explained, "Ella is 3 years old. It's time for her to learn how to ski. So she's taken two ski lessons so far, and she can actually fly down the bunny hill on her own. She looks like Frankenstein; it's hilarious. Dad is never more than 75 to 100 feet away with a defibrillator on his back. Ella doesn't know I'm there ... but we're not going to stop her from experiencing life and living." With his sensible attitude, Kevin breaks down the management of LQTS to a process of practical decision making that involves an objective assessment of the potential risks and the execution of appropriate accommodations: "The only difference with having a child with LQTS and not is making sure that you're doing those things that are responsible as an LQTS parent, ... so making sure that you know where the defibrillator is and that it's nearby, making sure that the medicine has been taken when it's supposed to be taken, and making sure that whatever activity you're doing is ok and is being accommodated for." Kevin does not view Ella as significantly limited by having LQTS, yet he stresses the need to be prepared to act at all times if she were to develop symptoms: "As far as restrict, we have not really restricted her in any way ... outside

of being too far away from where we can get to her and take action if we need to. We're very reluctant to lose line of sight of her just in case something happens.”

Other fathers expressed similar sentiments. “We work hard to avoid LQT restricting our children or change how we see them,” was one father’s response. Gary also expressed a perspective much the same as that conveyed by other fathers, stressing the need to let “kids be kids”: “You just let ‘em live, I mean, basically is what you’re doing. You just gotta watch the parameters that have been set up.” Collectively, fathers want to provide their children with the same opportunities for growth and development as those given to their peers for meaningful experiences that are not limited by their circumstances. A pragmatic approach is commonly adopted in managing children’s activities, using what is known about LQTS and associated triggers to make well-informed decisions and exercise good judgment.

### **Ongoing Challenges of Living with LQTS**

#### **Uncertainty**

Fathers generally view LQTS as a manageable condition in which the associated risks of cardiac events can be minimized via a proactive approach towards treatment and lifestyle considerations. Although individualized treatments can be highly effective in managing LQTS, fathers described different views on what they find to be most personally challenging about the diagnosis. All six of the fathers interviewed in the study spoke directly of emotional challenges associated with living with LQTS in their families, and six of the seven fathers who participated in the online survey referenced fear or uncertainty in their responses.

**“It’s always out there.”** Fathers described lingering worries associated with the potential risk of developing symptoms despite their children receiving effective treatment. Michael described the helpless feeling of knowing that despite taking every precautionary effort to protect his daughter from harm, there is no way to guarantee that something bad will not happen: “You feel, like, ... helpless because, you know, you’re watching, hoping it doesn’t happen, but it’s not like you can ... you can’t prevent it. You can do everything you can to try and prevent it, but it’s always out there like a time bomb. It could happen.”

While fathers’ emotional lives are not dominated by their fears or worries related to LQTS, most experience a persistent undercurrent of concern. The phrases, “It’s always in the back of your mind,” “It’s always in the back of my head,” and “It’s always in the background” were used several times throughout their interviews by four of the six fathers. Kevin’s daughter, who is now 3 years of age, was diagnosed at birth. When asked to describe how having a child with LQTS has affected him, he discussed the background hum of concern: “Well, there’s always that little ... that undercurrent that never stops, that thread of worry that is always there in the background cause you can’t ever stop thinking about it.”

**The watch for possibly developing symptoms.** Background stress is manifested behaviorally as fathers stay watchful for signs of potentially serious symptoms that they fear could develop. By remaining alert for potential signs of danger, fathers may feel better equipped to protect their children. Vigilance may increase during children’s participation in sporting events because of concerns regarding increased risks for cardiac events. Bob’s son, Ryan, was 10 years old when

he was diagnosed with LQTS, and already a competitive athlete. Bob recalled the nervous anticipation he experienced at his son's sporting events soon after his son's diagnosis. "We met with Dr. C, and she was very cautious of even letting him do anything from sports or activity-wise. So initially, when we had to deal with it with Ryan, I'm trying to think back, but you were much more aware of it, and much more on edge all the time when you had to sit through physical activities that he was participating in, and you knew you were sitting there with an AED and you don't know nothing about it right." The passage of time appears to moderate anxiety in the absence of LQTS-related symptoms but does not eliminate it. According to Bob, "It's still an unknown ... it's just in the back of your mind all the time."

**Overprotective or reasonably cautious?** While Kevin does not allow worries to limit his young daughter from enjoying a variety of activities and experiences, he is certain to always have his eyes on her. Young children are inherently energetic and naturally drawn to active play, and Kevin enjoys seeing his daughter engaged in play with her young peers while actively monitoring her. He said, "If she goes to the bouncy house, I'm watching her the whole time, and I've got the defibrillator in my hand ... I'm not going to stop her from going to the bouncy house, but I'm gonna make sure I get to her in less than 90 seconds." This observation raises a complex issue as to the level of parental vigilance that is justified and reasonable and whether a parent is at risk of becoming excessively cautious. Kevin's view seems to be based upon an acceptance of potential risk and a prudent approach to reducing those risks while allowing his daughter to remain active: "I'd say the only thing that's different is that we are, we're very reluctant to lose line of sight of her

just in case something happens ... so in that regard, we're a little bit more cautious than a lot of parents might be ... and I'm always watching for it, you know, if she drops I'm going to be on her."

Michael monitors his daughter vigilantly for potentially developing signs while attending her cheerleading events. Common symptoms, such as fatigue or becoming overheated, create ambiguity when the cause of those symptoms cannot be immediately determined. Michael described the heightened tension he experiences in balancing his response to potentially threatening symptoms while trying to maintain a reasonable judgment of the situation: "But we're always watching to make sure, you know, when she's cheering at the high school, then you're just watching to make sure she's looking alright, ... and she'll say, ... 'I don't feel right, I'm tired' ... and then I'm tense up on it, so little things that you would normally take for granted, and then when they say things, it's like whoaaa in the back of your mind; like well, she seems okay so I'm not going to panic yet ... so you're listening and watching for the symptoms and the signs all the time."

His daughter's diagnosis has undoubtedly shifted the way Michael processes information, most specifically in regard to health-related cues. He admits, "You just don't take a lot of things for granted that you used to take for granted, and you can't just say, 'Oh, she's fine,' because anything that they say now or they just don't feel well or something ... in the back of your mind, you're thinking, I've gotta keep an eye on this, get her some fluids, and make sure that they're doing all the things we're supposed to be doing, because ... it's out there ... it could happen, it's real, it's not something that you can just say, 'Oh, we'll be fine,' because that's not the case. It's

there, it's real ... you just can't ignore it." Michael described the change from his instinctive habit in the past to avoid overreacting to the often ambiguous physical symptoms that children display to his now much more guarded response to his daughter's behavior. Each symptom is mentally filtered as a potential signal of a more serious problem, and the risk is in allowing himself to become too comfortable and complacent: "It changes your life in that you're ... you constantly, I mean, my first reaction sometimes to things would be 'Oh, you'll be fine,' and I catch myself sometimes thinking that, and I'm like, you know, I can't do that, you know, I always have to be on guard. ... You can think they're going to be fine, but you still have to make sure they're doing the things to insure they're going to be fine, the medicines, the fluids, avoiding the stress, and make sure they've been eating their meals correctly and they're getting the right kind of foods and stuff like that. So, I'm always watching; so, it's always there."

**Divided attention.** As a regular spectator at his son's basketball games, Bob tries to distinguish innocuous signals from possible LQTS-related events, processing his child's behaviors through a mental filter attuned to potential threat. While he watches the game, Bob's attention is divided between his interest in his son's performance and his vigilant monitoring of suddenly developing symptoms. He stated, "You want him to do well, but you're watching the game with a different perspective of just watching at times how tired he is, and if he falls down and you're always like, okay, it's not is he going to get up because he hurt his ankle or whatever, it's like, is he going to get up? So, it has that kind of impact on you."

**“A little bit of panic.”** While very high levels of ongoing stress related to LQTS was generally not reported, fathers may experience rapidly escalating worries in certain circumstances. Worries may quickly surface, for instance, when events do not go as planned and there is no way of knowing with certainty whether children are safe and sound. For example, what happens when a child does not call or does not come home when expected? In the absence of an immediately obvious explanation, families may be inclined to assume that the very worst has happened. Bob recalled such a scenario occurring as recently as the evening before the interview with him took place. When his son did not return home by the expected time, both Bob and his wife became overwhelmed with panic that something had gone wrong. He related, “Yeah, it is constant, I mean it’s constant, I wouldn’t say panic, but it’s always in the back of your mind, and even like, last night ... cause Ryan still will exercise and go out to whatever gyms, so he was out and we knew he’d be there from 5:30 to 6:30, so we ordered out dinner, and it got to be 7:00, and she’s [his wife] trying to text him and call his cell phone and he didn’t answer. So then she’s freaking out, and we’re all starting to freak out, saying ‘Okay, did something happen?’, and it’s probably like anything else, but the first thing that comes to your mind is, ‘Oh, *shoot*. He was out exercising, and did something really happen?’ I’m driving out in the car to drive to the gym to see if he’s there, and then we’re like, ‘Oh, okay, he’s home,’ so we both feel it all the time. A little bit of a panic.”

**The challenge of comprehending the seriousness of uncertain risks.**

Fathers may also experience ongoing uncertainty about the seriousness of their children’s LQTS, particularly in the case in which their children do not exhibit

symptoms. One father described his greatest challenge as “living with the uncertainty of how seriously to take the condition.” Ted described similar challenges. His daughter, Tina, was diagnosed with LQTS at birth but did not experience any cardiac symptoms until the age of 15 years. Ted described his difficulty in determining the gravity of his daughter’s diagnosis when she had not experienced a single cardiac event for so many years. Ted recalled wondering whether the major procedures undergone by his daughter to prevent symptoms, including the implantation of an ICD and left cardiac sympathetic denervation surgery, were necessary. Ted recalled questioning the clinical judgment of the physician treating his daughter: “But over the years, she was asymptomatic and never had any issues ... We had all these kind of doubts as to whether the doctors really knew what they were doing, whether she really needed a device, whether she really had to go through all these issues her whole young life.” Children with ICDs must also contend with periodic testing to make sure the devices are operating properly. Ted described the additional stress he felt having to put his daughter through invasive test procedures while struggling with the uncertainty of even knowing if they were necessary: “When she’d go in and have things tested periodically, they’d ... cause her heart to go into arrhythmia in order to test the device, and just knowing that fact is kind of unsettling ... and then for years, though, we often wondered why she even needed to have this. She never had any symptoms of any kind, never, so we kept on ... wondering why she’s got to do this, and then ... she had to have another ICD implanted cause the battery was running low, and then still again, she never had any symptoms.” Several years later, however, Tina went into cardiac arrest, and Ted and his wife felt grateful that they “went

through all these procedures and all this heartache to make sure that she had a device to protect her.”

### **Traumatic Experiences**

Parents of children with LQTS may be at risk of traumatic stress in response to their children’s potentially life-threatening events. Ted was at home with his 15-year-old daughter when she went into cardiac arrest. He recalled, “When this event happened, you know, she said she was feeling kind of faint, she just felt funny. I sat her down, and I was going to take her blood pressure and she basically ... just kind of flat-lined right in my arms.” Witnessing his daughter’s heart stop is something from which he still has not recovered. Talking about the event during the interview was very difficult for Ted, even 3 years later. He admitted, “It’s something that I’ve been dealing with just as much as she has. I can’t get those moments out of my head. They’re permanently implanted there.” That frightening moment was followed by the further upset of witnessing his daughter receive a series of shocks from her ICD. Ted described the experience of being there with his daughter as she was transported in an ambulance, all the while helpless to comfort her: “I just left everything right there and got in the ambulance and just tried to console her, and you know ... the look on her face just before she knew what was going to happen to her was just really disturbing. She knew when her body was feeling a certain way that she was going to be shocked again.” Because she could not be stabilized, a helicopter was subsequently needed to transport Tina to a different hospital in which her cardiologist worked. In the helicopter, Tina was wrapped up “like a mummy,” and headphones were placed on her ears to muffle the sounds. Ted described again the feeling of powerlessness in

watching his daughter undergo repeated shocks virtually alone, with no one able to help her: “So, she’s going through this, and nobody can hear her going through it, and her mother is just watching from the back, and she can’t move to comfort her while she’s going and having these events, and the technicians don’t hear her, so she kind of went through all of that all over again totally by herself, knowing people were watching but they wouldn’t or couldn’t do anything about it.”

### **Daily Stress**

Some fathers reported stress associated with the additional burden that managing LQTS can place on everyday routines. When asked about the challenging aspects of LQTS, Gary described preparing for a school field trip: “But it’s tough ... I guess it all depends on the day that you’re having ... like you go away and you gotta make sure you pack everything you need just in case something happens. I mean, you’re carrying around a 15-pound object [AED] with you. It’s tough. ... It’s a heavy piece of equipment to put in a bag ... and you want to bring a camera, now you gotta pack a lunch ... now you got 22 pounds worth of stuff to get on the school bus. ... Just it adds up sometimes, and that’s when you think about it the most.” AEDs are recommended as part of an overall treatment program for individuals without an ICD. Carrying around an AED, however, may be not only a physical burden but also a source of stress for some fathers who view these precautions as mental reminders of what could go wrong.

Lifestyle changes intended to avoid known triggers may add stress to daily routines by restricting children from certain types of leisure activities. Gary, who enjoys the outdoors and spending time fishing with his children, described sometimes

feeling both physically and emotionally drained by those limitations: “It makes me a little bit more lethargic sometimes, like I’m thinking ... ah, it’s a hot day. We’ll just stay in the AC all day, you know ... like that sort of stuff, but it does, it wears on you some days, you know. Other days it don’t, you know, it all depends. Like today ... we could go out and do anything we wanted, but if you looked outside it was 102 [degrees]. I mean, me, I could still get on a boat 5 in the morning and fish ‘til 1... She would never, you know ... my boys and I would have been out there ‘til 3:30 in the afternoon and they’d have been fine ... but since the sun and prolonged exposure to the sun and how tired she gets, you know what I mean, there’s always a ritual that we try to stick to.”

Avoidance of certain activities may develop because of an inadvertent association between a child’s symptoms and a particular event, even when a medical basis is not apparent. Gary avoids certain activities because of his fears that they could trigger his daughter to show cardiac symptoms. His concerns stem from the circumstances of his daughter’s initial diagnosis, which surfaced after she was bitten by a tick, and his fear that another tick bite could be a catalyst to an even more severe episode. Gary stated, “I don’t want to bring her fishing. ... I don’t want to bring her through the field with ticks. ... I always think that’s what caused it and brought it on. ... What if it happens again?” Although the electrophysiologist treating his daughter says that this concern is not realistic, Gary said, “but we still keep her out of the woods.”

## **Marital Stress**

Fathers were asked to describe the impact of LQTS on their relationships with their spouses. Fathers described their relationships with their spouses as strong and supportive. In some cases, fathers feel that living with LQTS has even brought them closer together as a couple. Having a child with LQTS, however, requires parents to sort out the meaning of a complex diagnosis and the impact it will have upon their children. The task of managing and coping with a potentially life-threatening condition can place added stress upon a couple, as it poses a new challenge to the relationship and each partner's role. Uncertainty about how to best meet their child's needs may add stress to the relationship, and spouses may feel pressure to meet new demands. Bob described how the continual worry about how they are managing their children can be a source of added tension in their relationship: "I think it's just an added point of stress, right, in any relationship, of worrying ... that you're both doing what you're supposed to be doing with the kids and you're not pushing them too hard but something could happen." Bob admitted that he would be the one more likely to be questioned for pushing his children further. He said, "She [his wife] would find out about it and say, 'Well, what are you doing? You know you're not supposed to do that.' Yeah, probably a lot more stress around making sure that we both manage the daily lives in the way we should."

## **Finding Respite**

Parents of children with LQTS are more likely to watch over their children continuously and may be less inclined to allow their children to spend prolonged periods away from their homes and beyond their supervision. Finding time to

themselves may be a significant challenge for parents who fear leaving their children alone or in the care of others. Kevin, who is vigilant in monitoring his 3-year-old daughter, whom he never leaves out of his sight, discussed the challenge in finding a person whom they can trust to watch over their daughter in their absence: “I’ll tell you what’s really missing and the hardest part is finding a babysitter. Finding a babysitter that you can trust, that’s willing to take on the AED and that responsibility and that you trust actually leaving at home with your kid to be able to act on it.”

### **The Approach of Adolescence**

**The impact of restrictions on teens.** Fathers discussed additional challenges that may surface as children approach adolescence. Young children may adjust more easily to activity modifications adopted since an early age, while older children or adolescents may be more strongly impacted when suddenly restricted from an activity that plays an important part in their lives. Several fathers whose children were teenagers at the time when they were diagnosed received medical recommendations to discontinue high-intensity sports.

**Social pressures and the pressure to perform.** Children who play sports may also be subject to a more demanding set of performance expectations as they get older. Bob talked about the increased rigor of demands placed upon his son and daughter: “Our kids are allowed to play sports, but we always make sure that whoever is their coach understands what they have, and it’s not hard when they’re young, but as they get older, it becomes much more difficult. When you’re a coach of an athletic team and you’re run back and forth, run, run, run, and then if they’re not doing well with practice, they make the whole team do it. It’s like, ‘Listen, I’ll be there, and

she's not gonna do it,' or even him, when he was going through that." Older children and youth may also experience increased social pressures to participate on their school athletic teams. Bob's son was questioned by football coaches about his intentions to play, while implying their expectations that he would. Bob spoke about his son being approached by coaches at his school: "He's taller than me, he's probably 6'4", 6'5," so as he went through high school ... he had the pressure early on to play football. The coaches would walk by and say 'Well, aren't you going out for the team?'" Families have to step in and educate school staff about their children. Bob explained, "Elizabeth went to the same school, and because she knew who the coach was, and so we just told him, 'Listen, he can't ... so don't look at the kid and think you need to keep badgering him. He doesn't need to be badgered about this. He's not allowed to play."

**Increasing independence and fathers' protectiveness.** As children approach adolescence, fathers worry about their children's ability to manage themselves appropriately. Adolescents vie for increasing independence, spending more time with peers and less time under the supervision of their parents. They may be more prone to test limits, take risks, and adopt a carefree attitude without thinking about the consequences of their actions. Fathers worry about teens being responsible for their own health by making good lifestyle choices. Bob described his concerns as his children enter adolescence and may be exposed to potentially risky situations: "As it progresses to different things as they get to be different ages, as they're growing and just making sure that they understand that they have to take their medicine. Your relationship with them is more, I'll use 'cautious.' I don't know if cautious is the right

thing, but, you're more aware of maybe things that could go wrong, so you're a little more protective of them." Fathers want their children to be aware of the seriousness of their condition and to be appropriately cautious. They teach their children to be reasonably apprehensive and to act sensibly to avoid risky behavior. Michael said about his teenage daughter, "I don't want her to be fearless and thinking she can do whatever and not thinking about the things she needs to be worried about." Teenagers may also tend to assume they are invincible and immune to bad fortune. Michael worries that his daughter will fall into the mental habit of thinking that she is immune to danger and he therefore tries to instill in her a vigilant attitude: "Oh, it's not going to happen to me ... cause it doesn't, but it just takes the one time. So I feel like I gotta ... push 'em a little bit sometimes out of their comfort zone of just thinking it's not gonna happen, and making sure they're doing what they need to do."

Fathers may experience difficulty dealing with their teenagers' increasing freedom and their inability to monitor them and stop them from exercising risky behaviors. As Michael stated, "You're not with them in the car anymore, so you're always worried about it ... You just hope that she'll make the right choices, and fortunately, she's a very good girl and she makes good choices and she's done well and she knows the risks, but it's still there. You always worry about it." While Michael trusts his daughter's ability to make good decisions, he worries that her judgment may be compromised by peer pressure drawing her attention away from possible risks. Michael related, "She knows that, and I feel pretty confident, very confident in her because she assures me she understands the risk and stuff, but kids do at this age experimentation and things like that, and were she to do any kind of

alcohol or drugs or something, there's some drugs that could kill her, and she knows that, but ... I wouldn't want her to be in a situation where kids are ... 'C'mon' ... and pushing her and stuff, 'cause when I'm there I can jump in. I mean you can't do that."

**Being different and fitting in.** Kevin worries about how his decisions as a father will impact his 3-year-old daughter when she becomes old enough to reflect more deeply on her parents' choice to have a child who might be born with LQTS. Concern that she may feel different from her peers or have difficulty fitting in with others as she gets older, and possibly resent her parents for their decision, is something that Kevin thinks about but feels he can handle. He mused, "When she's older, now I've been around enough teenagers and seen the hormones go crazy with every child in the world to know that there is a possibility of resentment, and I know that kids, the worst things for kids is to be different, and there's gonna come a time where she's tagged as different because she's the kid with the AED, and that's gonna be a tough thing, but we'll get through it."

**Medical adherence.** Teenagers may also have a distrust of the information given to them by parents and their physicians. Ted's daughter has had an ICD since birth and suffered a cardiac arrest when she was 15 years old. She is now 18 years old and can be skeptical of medical information. Ted stated, "She has a distrust of the things that we tell her, even though we're not lying to her. We're being open with her. She just ... she basically distrusts all the information that is out there, and unless she can see proof of it in front of her of what we're saying, she's skeptical." This distrust can be problematic when children's adherence to their treatment is compromised by their fear that they are receiving incorrect information that could be harmful to

them. They may find contraindications in their own treatments based on their own Internet research and then fear that something is being overlooked by their physicians. “It’s caused a little bit of frustration, occasionally,” said Ted. “You know, we try to explain to her the importance of being compliant with her meds ... but then she starts reading how much she should be having, and then it’s like ‘I’m taking too much. The doctors are recommending that I take twice as much, [so] why am I taking twice as much?’”

### **Social Conundrums and Explaining LQTS to Others**

LQTS is a complex and uncommon condition. Most people have never heard of it. Families with children with LQTS will be confronted by a variety of situations in which they will have to explain their child’s condition to others. Fathers may find it difficult to explain their child’s condition to friends and acquaintances who as Bob described it, “Look at your child and say, ‘Well, what do you mean? They have what?’” Children with LQTS are often active and in good physical condition, and some may show no signs or symptoms of the condition. Challenges arise when trying to explain to others that one’s child has a condition that places him or her at risk for sudden death but is very manageable at the same time. Bob reflected on his own inclination to downplay the potential consequences of his children’s condition to others and considered his possible reasons for doing so: “But it’s still very hard, right? ... It’s hard because [her] physical appearance and [she’s] running around. You can say it to somebody, and they’re like, ‘Yeah, well, I don’t see anything. What’s the big deal? Yeah, it could be nothing.’ So, I probably find myself, if I’m talking to someone, I probably do that. I tell them, and then I might minimize it more than

anything by saying, 'Yeah, but I have it, and nothing's happened to me.' ... I tend to do that a fair amount." Bob considered that his tendency to downplay his children's condition could be a "defense mechanism," or a way to minimize his own worries. He continued, "I don't know if it's that reason, or it's maybe the other reason where you'd be sitting on the other side, and say, 'Well, what are you an idiot? If that's the possible implications, then why do you have her out here?' ... That does come into your mind often too ... People thinking the same thing: 'So, what are you a dummy that you have your kids out here doing this, if that's the case?'" In talking about when he started to develop this tendency, Bob brought up the possible fear of being evaluated negatively by family, friends, or acquaintances who may question his decisions regarding his child. Bob's son, who was already active in competitive sports when he was diagnosed, continued to play at a competitive level after his diagnosis. According to Bob, "He [his son] still played basketball, he still played soccer, he played very taxing physical sports, and that definitely was, and maybe that's where I started to do it, cause in one breath be talking about it, and that he can't do it, and then the possible implications, and then the next breath, you know, he's out there running himself to exhaustion trying to win the game and can just imagine people looking at me like, if I were you I don't think I'd have my son out there doing this."

### **Concerns Related to ICDs**

Several fathers interviewed and surveyed identified their greatest concerns as related to the use of ICDs. Fathers whose children have ICDs are concerned about potential complications, such the device not working properly, long-term

maintenance of the device, and their children requiring lead removal. The costs of ICDs are very high, and fathers worry about children being able to maintain insurance to cover the costs of ICD replacement once they reach adulthood. Deciding whether or not to have their children receive an ICD is a difficult choice for fathers. When asked to describe his most difficult decision regarding his child's condition, one father replied, "The decision to use or not to use an ICD has been the most difficult." His greatest concern is that his son will have a fatal event without the use of an ICD.

Fathers worry about the future and that their children will one day demonstrate a need for an ICD. Kevin's daughter is only 3 years old and is treated with beta-blockers. She has never had a cardiac symptom or event. Kevin's wife was diagnosed with LQTS and has an ICD. Kevin described one of his greatest concerns as his daughter possibly needing an ICD one day. "The other [concern]," he added, "is if she were to have to get an ICD. I'm holding off on that part as long as possible. ... That's going to be a very, very long debate and don't exactly know where we're going to end up on that one, but that debate, hurdle, we have coming down the road is at the point in time where her EP [electrophysiologist] says she needs one ... but when that time comes ... that'll be a group decision and one that we do exhaust the research on and ... it'll be the last option."

### **Fathers' Coping**

#### **Knowledge and Education**

*"We can deal with what we know. What we can't deal with is what we don't know,"* said Kevin emphasizing the importance of staying informed about LQTS. The diagnosis of LQTS is scary and confusing to fathers who have never heard of it and

do not understand it. In response to their concerns, families actively seek to educate themselves. Fathers want to know what they are dealing with so that they can take the appropriate steps for their children. Many fathers discussed the importance of knowledge in regard to managing LQTS. They said that knowledge allows them to gain a sense of control in managing their children's LQTS by following the recommended guidelines and precautions. According to Kevin, "There's so much of it out there that isn't known, and if you at least know you can do something about it, you can avoid certain activities. You can avoid certain situations. You can take the beta-blockers. You can have an AED." Parents of children with LQTS face many challenging decisions involving their children's medical treatments and lifestyle, and fathers use knowledge to make informed decisions. Kevin is an analytic thinker who uses knowledge about LQTS and the associated risks to make informed and objective decisions. When asked about whether he worries about his daughter when she is away during the day attending her preschool class, he responded, "I guess I'm really good at compartmentalizing stuff. It's the additional risk of having something happen while she's at school is acceptable to me to give her the benefit of going to school and being around the other kids and learning." Accepting the inherent risks that may exist in any situation, Kevin sifts through his emotions to better objectively engage in a decision-making process.

### **Reframing and Seeing Through a Positive Lens**

Fathers described various ways in which they cope and adapt to living with children with LQTS. Michael and Kevin maintain a positive outlook about their children's well-being by thinking differently about their situation. Michael views

himself as fortunate to have discovered that his daughter has LQTS and to have the knowledge that he needs to protect his wife and daughter. The tragic circumstance would be to not know of her underlying condition and then for something bad to happen. He explained, “You want to protect your kids and your wife and you don’t want any of this to happen, but I almost feel it’s good to know. I felt like I’m lucky that I found this out so I would know, so that if we’re doing the precautionary things now, we don’t have to be one of those that finds out the very tragic way. So, I guess I felt lucky that we were ... that we found out in a nonthreatening way through normal testing and stuff where nobody had to go through a sudden incident where they almost died or died.”

The potential risks to their children are always in the back of their minds, as fathers put it, and these background fears can be a source of ongoing stress. Kevin knows that the development of symptoms without warning is a possibility and described an undercurrent of worry that never goes away. Although these concerns have the potential to elicit fear, Kevin sees this ever-present thread of worry as functional: “It’s a safety net. I think it’s there for a reason ... to keep people safe, to keep people aware, to make sure that you remember where the defibrillator is, to make sure that you’re doing the best.” By reframing worrying thoughts as useful data, Kevin is able to channel stressful emotions by taking purposeful action.

### **Acceptance**

Adjusting to the idea that their children are at risk for life-threatening cardiac events is a challenge which parents face when their children are diagnosed with LQTS. In considering the worst possible outcome, the sudden death of his child,

Kevin has no regrets and accepts what he can and cannot control in regard to his daughter's condition. "If the worst happens tomorrow and I lost her, it would have all been worth it." said Kevin. ... "The way I look at it is she's got this condition and we're doing everything that can be done to address it and keep her safe ... The fact that we know is such an advantage, the fact that we know and that we can do something about it to stack the odds in our favor so much more than they would have been if we didn't know and weren't doing anything about it. The bottom line is anybody walking down the street can get hit by a bus, and it's worth the chance. It's part of the total package, and it's worth every bit of it." Gary talked about learning to be flexible enough to adapt to changes and accept those changes as part of a "new normal." In discussing his adjustment to his daughter's diagnosis, he shared, "Once you realize it's normal for her ... you just gotta roll with it."

The idea that many aspects of life are beyond human control was discussed by Amir, whose beliefs about God and fate are central to his way of coping. Amir endured extreme duress at the time of his daughter's birth and then had the task of negotiating one tremendous obstacle after another to obtain his daughter's treatment. When discussing how he has coped with these severe circumstances, Amir reflected on his belief in God, which has shaped both his perspective and response to the adversity he has faced: "God pitched a very difficult test against us, but He also gave us wisdom and luck and many factors to deal with this difficult test. So, eventually my faith in God was restored and it got more strong, and now I feel that always the problems are pitched by God to test us in mysterious ways. So, the strategy of believing in God and believing that what he does is best for us human

beings, He's watching us, He has given us different [tests], but He has also given us different skill sets. So yeah, I believe in destiny and fate; whatever is written is the plan. It's all a plan of God. I tend to cope with all this stress and all this uncertainty that there is an entity, a heavenly entity, who has all the fate and all whatever is happening with us in His hands."

### **Adjusting Expectations and Reprioritizing**

Once his son was diagnosed with LQTS, Bob reflected on the difficulty he had adjusting his expectations, particularly as they related to his son's involvement in sports. Bob described how he would recognize his son behaving cautiously as an athlete, and then would push him harder: "So when you saw him and you saw the same group of kids that he would participate with and he was more cautious from once he was diagnosed, a lot more cautious ... where I would still push him to say, 'What are you doing? You can do this.'" Bob believes that adjusting to this new set of circumstances may have been harder for him than for his son: "I probably had a harder time adjusting to it than Ryan did and coming to the realization. It took me a lot longer to understand this, that this isn't that important." Bob discussed being raised in a family of competitive athletes and growing up himself playing sports at a competitive level throughout college. Bob was diagnosed with LQTS as an adult after his son's condition was discovered and had never experienced any symptoms while growing up. He stated, "Remember, he [Ryan] was still pretty competitive and I'm pretty competitive, so it was more pushing him at that with those same expectations, or as the expectations or the reality had changed. So, that's not going to happen, so what are you doing this for?"

Bob described how having children with LQTS shifted his focus so that certain ideals which had once been most important to him no longer were. Over time, his son's ability to participate and compete with others without the pressure of cutthroat standards of performance became more important to Bob. His priorities were put into a broader perspective by his son's physicians and staff. He conceded, "You know, this whole period of their lives where they're pushed to be active here, there, and everywhere and compete at everything is a very small portion of their life." LQTS caused Bob to refocus on new priorities. While he still may view involvement in athletic activity as having a positive role in children's lives, it is no longer pursued with the same drive to excel or need to be the best in a sport. He summed up by saying, "So it did make you focus differently. I mean, the focus was on academics, the focus was on different things to make sure that they pushed themselves academically and things of that nature." Bob pointed out the unintended benefit that refocusing his priorities and, in effect, removing pressure to excel at sports had on his relationship with his son: "It kind of took the edge off."

When fathers are faced with the seriousness of their children's life-threatening condition, their perspective on life itself changes. Kevin talked about how living with LQTS has affected both his wife's and his own appreciation of life itself: "We try not to sweat the small stuff as much, and we really appreciate life. I mean, we appreciate every day we get because there's always that little background undercurrent of something bad could happen and they may not be there, and I do at some level recognize more strongly now since Adrienne's diagnosis and since Ella's both the value of every single day."

Amir described how his role as a father and his priorities changed with the birth of his daughter with LQTS: “I am like many a person who cannot compromise on his sleep, but for this child, I compromise on everything, even if it’s my sleep, financial matters, or commitments, like coffee and friends. So yeah, we have to compromise a lot to go to her medical appointments, and we have to make a lot of changes. For me, my job is a secondary priority now, if some issue with her, sending her data out or talking to doctors, everything is a secondary priority now. She is a foremost priority now.”

### **Sharing Their Story**

During their interviews, several fathers said that they were glad to have been given the opportunity to share their stories with others and hoped that by doing so others might benefit. Most fathers are involved in different LQTS groups in their community or on social media, where they have also talked about their stories. Fathers want something good to come out of the experiences they have had as parents of children with LQTS and to prevent, if possible, tragedy befalling others: Michael shared, “ [I] share our story with people I come in contact with and people at work, and just let them know about it, that it’s out there, and that other people should get checked as well, and that you just never know, and I’ll see stories where kids have died suddenly, and I just, right away, I feel terrible for them, the parents, because I think that they didn’t get that warning, and I almost feel blessed that we got the warning, but I feel so bad, and we do a lot with Simon’s Fund and trying to help them be successful, because I’m glad that we’re not one of the parents that found out through tragic ways ... I also feel bad for those that do have to find out that way, so I

share our story and I always tell people about what my daughter's condition is, and just what we're watching for and stuff and just letting them know, that it's out there ... and for people to be aware more than anything else."

### **Resources/ Supports**

Fathers identified their physicians as the resources most helpful to them in living with LQTS in their families. Fathers described their physicians as accessible, knowledgeable, caring, and supportive. Several fathers discussed their satisfaction with being able to communicate frequently with their physicians through e-mail and receiving prompt replies. One father said the following about his child's pediatric electrophysiologist: "We talk through all our concerns. The doctor seems very open to input. He defers to those better placed to help. He has been very willing to work with us by e-mail, which is helpful." Fathers speak highly of their physicians, who show genuine concern for their families. One father said, "We have the best heart doctor [who] treats us like family and the whole staff is great and has taken great care of our son, Louis. I think she has saved his life." Expressing a similar sentiment, another father commented about his physician: "Excellent- wonderful professionals who really care and give us a great deal of hope." Kevin described his positive experience with his daughter's electrophysiologist, who takes the time to help them to navigate through various decision points: "You know we are very open, and one of the things we like about Dr. B is that she's very accessible. She spends a lot of time and clearly cares about her patients, and we talk about what are the boundaries, so you've always got to assume the worst, and we talk to Dr. B about what makes sense."

Other than their physicians, fathers relied mostly upon their wives as sources of support, although the fathers were even more likely to deal with issues on their own. Several fathers felt that they were able to handle the stresses related to LQTS without needing additional support. Several fathers were familiar with social media groups associated with LQTS and heart conditions, while their level of participation in these groups varied, and some fathers described themselves as more passive members of online groups. Fathers appreciated the practical information provided on websites, such as those providing lists of QT medications. Ted's bigger concern is finding persons who have had experiences similar to those of his daughter with whom she can connect and relate: "More importantly to me, in that respect is I like to see that my daughter can interact with people who are going through the same thing that she's going through, not so much how the family's coping. I mean, everybody's gonna have a little different mechanism to deal with that. I'm not trying to minimize that, but to me what's more important is for the people that have the condition to be able to reach out and get support from somebody else that has it. I mean this has been the biggest struggle for Tina is finding somebody to relate to, talking to them ... I have problem A, how do you deal with problem A? You know, I have problem B, how do you deal with problem B?, and they can share and exchange. So, it's kind of like, to me, well, while it's important to have the parent support groups, it's even more important to have support groups with peers of people who have the condition."

Amir was grateful to physicians in the United States who provided medical, logistical, and emotional support to his family in obtaining treatment abroad for his daughter that was unavailable in his country. He explained, "The medical

professionals in the United States, they knew that we were in a very difficult predicament, so we used to see in their eyes that they have pity for us. They felt in their hearts what we were going through. So yes, I know that they tried to help us as human beings as well. They tried to restore our faith. So yes, they were more than doctors, they were more than healthcare professionals for us. They went out of their way to help us in different ways, not only medically, but logistically, emotionally. Yes, they have helped us in various ways.”

## **Chapter 5**

### **Discussion**

#### **Summary of Findings**

According to participants in this study, fathers experience fear, worry, and confusion when their children are initially diagnosed with LQTS. Initial fears are moderated over time with increasing knowledge about LQTS while ongoing worries about the uncertain threat of LQTS-related symptoms remain. The findings of this study suggest that fathers play an active role in their children's lives and seek normalcy by accepting new family routines into daily life. Fathers recognize the seriousness of their children's condition but continue to view their children and their relationships with their children as unchanged in a number of important ways. The level of psychosocial stress that fathers experience over time may vary according to a number of biological, psychological, and social variables, and fathers appeared to use a range of strategies to manage stress associated with their children's LQTS.

#### **Fathers' Involvement and Influence**

While each family is different in the division of its roles and responsibilities, fathers may be actively involved in care-taking roles traditionally assumed as the primary responsibility of mothers. Fathers may monitor their children's activities, medication, and diet, and those who work from home may assume many primary-care roles. Fathers want to maintain as normal a lifestyle as possible in which LQTS is not viewed as a barrier to children's quality of life. The findings of this study suggest that fathers appreciate being involved in decision-making processes with their

spouses and healthcare providers regarding children's treatment and activities, with thoughtful consideration to children's talents and interests.

### **Striking a Balance**

The fear of their children developing cardiac symptoms leads to heightened vigilance, and fathers protectively monitor their children for possible signs of LQTS. Fathers described themselves as having become more cautious since the LQTS diagnosis, while seeking a balance in allowing "kids to be kids." Fathers spend time with their children in athletic and leisure pursuits while making accommodations to meet their children's needs. With their older children and teenagers with LQTS, who spend more time with peers and beyond parent supervision, fathers strongly encourage them to take a reasonably cautious approach in evaluating the potential risks of their choices and self-managing their behaviors. When children are diagnosed with LQTS at a young age, fathers are more likely to deliberately shape their children's interests towards activities that pose less risk for triggering cardiac events.

### **Factors Influencing Fathers' Adjustment**

The findings of this study suggest that while fathers share many similar concerns regarding their children with LQTS, each family faces its own unique set of challenges and strategies for managing those challenges. A number of variables were described that may influence fathers' adjustment, including LQTS-related factors, uncertainty and daily stressors, and fathers' cognitive processes and coping methods.

**LQTS-related factors.** LQTS is not one homogeneous syndrome but consists of many related syndromes with varied genetic and clinical features (Berul, 2008).

Children with LQTS vary widely in terms of frequency and severity of cardiac events, treatment plans, and activity restrictions. Some families have more than one child with LQTS and have relatives who have been affected by LQTS, including some who have died from cardiac arrest. Alternately, some families have children diagnosed with LQTS who exhibit no syndrome-related symptoms. Because families with LQTS may have widely varying experiences, fathers who have children with LQTS are also likely to have their own unique responses. While fathers may empathize with other families affected by LQTS, they may perceive their own experiences to be very different from those of others. In this small sample of participants, fathers whose children were asymptomatic or had experienced a single syncopal episode and were responding positively to their treatment generally reported lower levels of ongoing distress. Some fathers in this study had children who had undergone one or more cardiac arrests and required more invasive procedures, including ICD implantation and surgical intervention. Fathers whose children experienced one or more significant cardiac events appeared to be at greater risk for increased stress related to traumatic experiences, worries about possibly developing symptoms, and concerns about the impact of LQTS on their children's future.

**Dimensions of uncertainty related to LQTS.** The findings of this study suggest that most, if not all, fathers, regardless of the course of their children's symptoms, experience ongoing uncertainty and related stress. Various aspects of the illness experience underlie the phenomena of uncertainty and affect psychosocial adaptation (McCormick, 2002). Fathers described the following multiple dimensions of uncertainty associated with LQTS.

*Unpredictability and “the time bomb.”* Underlying the concerns of many fathers is their inability to know for certain if their children will experience cardiac problems. The wide-reaching clinical spectrum that defines LQTS and includes the possibility of sudden death does not allow fathers to predict outcomes for their children. While significant advances have been made in the clinical and genetic underpinnings of LQTS, the development of truly robust prediction algorithms remains difficult (Abrams & MacRae, 2014). The findings of this study suggest that the possible threat of future symptoms is always in the back of the fathers’ minds despite taking proactive steps to reduce the risk.

*Ambiguity and uncertain meanings.* Ambiguity arises when the meaning of an event or circumstance can be interpreted in more than one way (McCormick, 2002). Fathers experience uncertainty in ambiguous situations related to their children’s LQTS, beginning with diagnosis and with ongoing management. Although genetic testing may provide a confirmation of diagnosis, the early clinical stages of diagnosis can be challenging and may elicit ambiguous findings. Overlap may exist on ECG findings between the normal population and genetically affected individuals with no or mild QT prolongation, thus leading to inconclusive findings for children suspected of having LQTS. Additionally, fathers face uncertainty when the cause of children’s ambiguous symptoms cannot be immediately determined. Fathers are more attuned to children’s vague complaints of being tired or just not “feeling right,” which contribute to stress when perceived as a possible sign of something more serious.

***Complexity and seeking understanding.*** The medical complexity of LQTS makes the condition difficult for fathers to understand, thereby contributing to uncertainty regarding diagnosis, prognosis, and treatment. Diagnosis, prognosis, and treatment for LQTS depend on a complex array of historical, clinical, and genetic factors. Determining the level of risk for future cardiac events remains a considerable challenge and depends on careful evaluation by an expert clinician (Abrams & MacRae, 2014). Fathers may question whether more aggressive treatment procedures (i.e., pacemaker implantation, ICD placement, Left Cardiac Sympathetic Denervation) are necessary for their otherwise healthy children while also worry about the risks of choosing less invasive treatments. Even when treatments are physician recommended, fathers may still question if they are making the right decision for their children and may seek other opinions.

***Daily management and related stress.*** LQTS is generally viewed by fathers as a potentially threatening but manageable condition that can be effectively controlled with appropriate treatment and lifestyle modifications. The findings of this study suggest that fathers vary in terms of the amount of daily stress they experience associated with their children's LQTS. Some fathers reported greater stress associated with the daily management of LQTS, including frequent monitoring of children's activity, monitoring of medical and dietary compliance, limiting activity and avoiding triggers, and using such precautions as keeping AEDs on hand. Other fathers seemed more comfortably habituated to the daily tasks involved in managing their children's condition and did not perceive them as burdensome. Fathers with more stress seemed more prone to worry about their children and more likely to

perceive daily tasks as worrisome reminders of their children's condition. Fathers working from home may experience the additional burden of dividing their attention between their work responsibilities and monitoring their children at home.

Having a child with LQTS appears to strengthen family bonds, and fathers rely upon their wives for support. The additional stress, however, of having a child with a potentially life-threatening condition can put a strain on family and marital relationships, and fathers may question if they are adequately managing their children's condition. Fathers may be uncomfortable leaving their young children, in particular, under the supervision of other adults, such as childcare workers or babysitters, possibly resulting in less time for self-care and respite.

### **Significance of the Findings**

This study adds to the emerging number of studies examining the psychosocial adjustment of parents whose children have been diagnosed with LQTS. Existing research suggests that many parents of children with LQTS have difficulty adjusting to their children's new diagnosis of LQTS and that parents may experience ongoing worries over time (Hendriks et al., 2005). Previous studies examining parental adjustment to LQTS have examined mothers' responses or have examined parental adjustment without separation of mothers' and fathers' individual experiences. While fathers have historically been underrepresented in pediatric research, more attention has been given to the paternal role in managing children's chronic illness and adopting a holistic approach to treatment. This study is the first to specifically examine the experiences of fathers of children with LQTS and to describe their responses. According to the participants in this study, fathers are involved in the

ongoing management of LQTS and its impact on their children's lives, and these findings support a family-oriented approach to LQTS management informed by an understanding of fathers' unique concerns, coping, and adjustment.

### **Findings as Related to Previous Work in This Area**

These findings are considered in respect to previous studies investigating psychosocial factors impacting parents of children with LQTS. In examining current findings within the context of related research, one should note that the present study is the first in the literature to specifically examine the responses of fathers of children with LQTS. Both Gonzales (2009) and Burns-Pentecost (2013) reported exclusively on the responses of mothers whose children were diagnosed with LQTS. Hendriks et al. (2005) reported primarily the responses of parental couples of children with LQTS without separation of mothers' and fathers' responses. Farnsworth et al. (2006) analyzed the responses of adult participants in a primary pilot study who were diagnosed with LQTS and were also parents of children with LQTS. The study, however, provided no information regarding the gender of the participants. While several common themes were found across all studies, some varying responses were indicated.

A pattern of fear and worry among parents regarding their children's vulnerability to suddenly developing symptoms of LQTS, particularly close to the time of the initial diagnosis, was revealed across studies (Burns-Pentecost, 2013; Farnsworth et al., 2006; Gonzales, 2009; Hendriks et al., 2005). Such factors as increased knowledge and the passage of time moderate some parents' fears, while the continuous threat of possibly developing symptoms elicits an undercurrent of worry

(Burns-Pentecost, 2013; Farnsworth et al., 2006; Gonzales, 2009; Hendriks et al., 2005). The uncertain outcomes associated with this potentially life-threatening syndrome were described by fathers as “fear of the unknown,” similar to descriptions given by parents in prior studies (Burns-Pentecost, 2013; Farnsworth et al., 2006; Gonzales, 2009; Hendriks et al., 2005). The need to continually monitor their children for signs and symptoms of LQTS was another trend in parents’ responses across studies. Regarding children’s medical treatment, many parents reported a sense of uncertainty in determining the appropriate treatment for their children and also questioned the efficacy of chosen treatments. Previous findings indicate that parents manage fear and worries by taking direct steps to reduce or avoid potential triggers through medication management, lifestyle choices, and ongoing self-education and educating others in their community (Farnsworth et al., 2006). The current findings reveal that fathers want their children to safely continue with normal activities to the degree possible while monitoring their children’s activities to avoid potential triggers of cardiac events. They are present during their children’s athletic events and take precautionary measures, such as carrying AEDs on trips, outings, and sports events and practice trainings. They educate and train others who work with their children, providing information about LQTS and teaching others how to use AEDs.

Another pattern revealed across parental responses to their children’s LQTS was in regard to the impact of medical treatment and lifestyle decisions on children’s quality of life (Farnsworth et al., 2006; Gonzales, 2009; Hendriks et al., 2005). The need to maintain a healthy balance of normalcy and caution in considering lifestyle modifications for their children was expressed by many parents advocating for a

decision-making process guided by knowledge rather than fear (Farnsworth et al., 2006; Gonzales, 2009). Parents reported worries about possible side effects of medications, as well as about the potential complications resulting from ICD placement. Having access to knowledgeable healthcare providers in the ongoing medical management of their children was commonly expressed as a need by parents, particularly given the need to avoid certain medications associated with QT prolongation. Regarding their children's futures, parents worried that medical treatments and lifestyle restrictions would have an adverse impact on their children's careers, marriages, and families (Farnsworth et al., 2006; Hendriks et al., 2005). Another common concern of parents was the anticipated stigmatization their children might endure due to being viewed as different from their peers.

The approach of adolescence was a source of concern for parents across studies. They anticipated a diminished ability to control and manage their children's behavior and compliance with medical treatment plans (Farnsworth et al., 2006; Gonzales, 2009; Hendriks et al., 2005). As a result, many parents expressed a wish to instill a sense of responsibility, self-management, and reasonable caution in their children with their increasing levels of independence. Unlike fathers in this study, Gonzales (2009) found that some mothers of children with LQTS reported significant stress resulting from teenagers' increasing rebelliousness and possible medical noncompliance, while only one father in the present study reported his daughter's growing skepticism regarding her medical treatment. Of note, however, is that mothers were more likely to report concerns regarding their sons' increasing defiance, while all of the six fathers interviewed for the current study had only daughters with

LQTS who were 18 years or younger. Furthermore, a few mothers had hopes that their teenage children would be persuaded to receive ICDs because of their ongoing fears of developing symptoms, while no fathers indicated similar views regarding ICD treatment for their children. Although beyond the scope of this study, further research in this area is warranted to investigate whether mothers are more likely than fathers to seek more protective treatment options for their teenage children and whether parental gender or child gender influences children's compliance with medical and lifestyle recommendations. Further study is needed to examine how couples of children with LQTS make decisions when they hold different views regarding children's treatment and lifestyle adjustments.

Gonzales (2009) found that several mothers expressed feelings of disappointment and guilt regarding their children, particularly their sons, being restricted from competitive sports activities. Burns-Pentacost (2013) also reported that a minority of mothers expressed frustration in regard to the restrictions placed on their sons' activities. Fathers, as a whole, were less likely to express these sentiments, while, as noted, one father expressed considerable difficulty adjusting his expectations regarding his son's reduced level of competitiveness following the discovery of LQTS. Otherwise, all of the fathers interviewed had only daughters with LQTS who were 18 years of age or younger at the time of the study. Further study, however, is needed to investigate how sports restrictions may influence children and families differently according to gender.

Burns-Pentacost (2013) examined the responses of participants who posted to an LQTS online user group related to "new diagnosis." Of note is that all nine

participants were mothers, suggesting that mothers may be more likely than fathers to seek out online user groups in response to their children's diagnosis. Several mothers participating in this study described an absence of emotional support and understanding from their husbands, family, and community. In contrast, fathers denied the need for emotional support and felt they could handle their own emotional needs sufficiently or rely upon their wives. Mothers appeared much more likely than fathers to seek connection and emotional support through the utilization of Internet-based LQTS groups.

Mothers, as compared to fathers, seemed more likely to express initial negative encounters with medical professionals related to their children's diagnosis, along with feelings of being invalidated, discounted, or isolated (Burns-Pentacost, 2013; Gonzales, 2009). Despite this trend, several mothers expressed a more positive relationship with healthcare workers over time (Gonzales, 2009). Whether more mothers than fathers were likely to experience dissatisfying medical encounters because mothers were more often attending children's medical appointments is unclear.

### **Relevance of the Study to the Theory and Practice of Psychology**

#### **Applying a Risk-Resistance Adaptation Model to Fathers' Experiences**

Fathers' experiences can be examined within the context of a broader theoretical model to identify factors that may influence paternal adjustment to children's LQTS and that mediate and moderate the relation between illness-related stress and adjustment (Hocking & Lochman, 2005). Risk-resistance models have been developed to explain adjustment to childhood chronic illness by studying the various

factors that either place individuals at greater risk for adjustment difficulties or serve as protective factors in response to stressors (Hocking & Lochman, 2005; Wallander & Varni, 1989). Applying a risk-resistance model to fathers' experiences provides a framework to understand the relationship between various biological, cognitive, and social variables and fathers' adaptational processes.

### **Risk Factors Associated with Fathers' Psychosocial Stress**

**Presence of life-threatening events.** LQTS is a heterogeneous syndrome that affects individuals differently in type and severity. Some fathers who participated in this study had children who displayed no symptoms, and others had children who had experienced one or more cardiac events, including sudden cardiac arrests. Some children with LQTS remain asymptomatic throughout their lives while other children may experience recurrent syncope, seizures, cardiac arrest, or sudden death. Children have different treatment and maintenance needs over time. Ongoing concerns about their children's well-being were expressed by nearly all of the fathers in this study because of the potential threat of possibly developing symptoms, while absence of cardiac events over time appeared to moderate these concerns. Fathers whose children had displayed more significant symptoms of LQTS (i.e., cardiac arrest) experienced traumatic memories and expressed more concern about the impact of LQTS on their children's emotional well-being, careers, and future autonomy than did fathers of children with LQTS who displayed less significant or no symptoms. More significant symptoms of LQTS were more likely to result in children's absence from school for extended periods, thus requiring increased care-taking responsibilities and potentially impacting fathers working from home.

**Uncertainty and associated stress.** Fathers who experience the most uncertainty related to their children's LQTS may report higher levels of stress. As previously described, uncertainty is present from the time of the initial diagnosis of LQTS throughout treatment along various dimensions. According to Mishel (1984), uncertain situations are those perceived as unpredictable, ambiguous, complex, and unfamiliar. The diagnosis of LQTS often comes as a surprise and shock to families unfamiliar with this potentially life-threatening syndrome. LQTS is associated with a wide-ranging clinical spectrum, and fathers who experience their children's LQTS as highly unpredictable and ambiguous in nature may experience significant stress. A lack of adequate information on LQTS may further contribute to a sense of the complexity of this condition and interfere with fathers' confidence in their ability to make the best decisions for their children regarding their treatment and lifestyle.

**Restrictions on children's activity.** Fathers value the benefits of children's participation in sports and recreational activity, and they support and guide their children's interests. Many children with LQTS can safely participate in many forms of recreational activity while children with a history of cardiac arrest, suspected LQTS-related syncope, or prolonged QT interval may be more susceptible to cardiac events during sports and vigorous activity (Maron et al., 2004). Fathers whose children face stricter lifestyle limitations may themselves be at increased risk for stress. Activity restrictions may disrupt everyday family routines, and fathers may become more cautious and guarded in monitoring their children. Daily avoidance of triggers may place limits on leisure, sports, and recreation, placing additional strain on fathers. Fathers who value the role of competitive sports in their children's lives

may find themselves confronted with a need to rethink their priorities and adjust their expectations.

Schools may restrict children with LQTS from competitive activities, physical fitness tests, or certain components of physical education courses, depending upon the particular policies held by their school district or state. Social and emotional consequences may ensue for children who, as a result of such restrictions, are excluded from normal activity and may feel ostracized by their peers. Fathers may experience additional strain in managing both the physical and psychological aspects of LQTS on their children's development.

**Child's age and the approach of adolescence.** The child's age at the time of diagnosis may influence the way in which LQTS impacts the entire family. When LQTS is discovered during a child's adolescence, established lifestyle routines and activities may need to be modified. Children and teenagers who are already actively involved in competitive sports activities may be restricted from such activities, causing a significant disruption in children's lifestyle. Families whose children are very young when diagnosed with LQTS are likely to direct their children towards interests and activities that pose less risk to them, and children may adjust more easily to lifestyle habits that they are accustomed to from a young age. Nevertheless, as children approach adolescence, families are likely to experience additional stressors that arise as their children seek growing independence. Fathers expressed concerns regarding their teenagers' ability to use good judgment and avoid risky behavior and maintain compliance with their medications.

## **Resistance Factors Contributing to Fathers' Adjustment to LQTS**

**Fathers' appraisals of stress and uncertainty.** The stress and coping model (Lazarus & Folkman, 1984) suggests that a key factor in fathers' coping and adjustment is their appraisal of stressful events associated with their children's LQTS. Illness-related events may be appraised according to the extent to which events are perceived as threatening, harmful, controllable, and challenging (Lazarus & Folkman, 1984; Riley & Park, 2014). Uncertainty is a common component of illness-related events and is appraised as threatening when the stakes are high, such as with a potentially life-threatening childhood condition (Mishel, 1988). The complex and wide-ranging clinical spectrum of LQTS generates a state of uncertainty in fathers who may experience the most intense fear when the diagnosis is first discovered and least understood.

Ongoing uncertainty associated with the potential risk of cardiac events is experienced by fathers, while their appraisals of LQTS may change over time (Mishel, 1990). Several factors appeared to moderate fathers' perceptions of uncertainty associated with their children's LQTS. One of the most significant factors impacting fathers' perceptions appeared to be their growing knowledge and understanding of LQTS. As families acquired a better understanding of LQTS and its effective treatment, most fathers viewed their children's condition as more manageable and less threatening. Additionally, the passage of time appeared to be a significant factor in moderating fathers' appraisals, with the absence of developing symptoms over time reducing the perception of looming threat and increasing a perception of control over their children's condition.

While time and knowledge appear to alter perceptions of children's vulnerability to threat and associated fears, continual uncertainty regarding the potential impact of LQTS arises from the inability to absolutely eliminate the risks associated with LQTS. According to the reconceptualization of uncertainty in illness theory (RUIT), one's perception of uncertainty as threatening may gradually change over time as individuals adopt new perspectives and identify beneficial aspects of uncertainty (Mishel, 1990). Some fathers positively interpret their uncertainty of potential cardiac events as a beneficial reminder to take proper precautions. Additionally, positive appraisals of uncertainty can be facilitated by fathers who accept ambiguity and unpredictability as conditions of life and accept a world in which absolute certainty is unattainable (Lin, Yeh, & Mishel, 2010). In Mishel's theory (1999), the process of accepting a positive view of uncertainty is labeled as growth through uncertainty. Fathers who accept the uncertainty of living with LQTS may demonstrate this growth by becoming more motivated to appreciate and make the most of each day (Lin et al., 2010). Some fathers recognized uncertainty as an aspect of living with LQTS but viewed their families as able to maintain as normal a lifestyle as possible that was not limited by LQTS.

Religious and spiritual beliefs may also influence fathers' appraisals of illness-related events and the uncertainty associated with illness. Strong beliefs in a higher being may facilitate greater acceptance of the uncertainties of life as fate in the hands of a higher god or spirit (Karekla & Constantinou, 2010). Religious beliefs may also facilitate perceptions of events associated with illness as having a purpose or meaning that may not be immediately apparent to one's comprehension. Fathers who

practice religious and spiritual beliefs may accept a limited perception of control in regard to LQTS within the context of a fate that is controlled by a higher being.

**Fathers' ways of coping.** According to the transactional model of stress and coping, appraisals of events do not directly influence psychological adjustment but are mediated by coping, and choice of coping is influenced by one's appraisal of events (Lazarus & Folkman, 1984). As noted, fathers' appraisals of LQTS-related events may shift over time and influence their selection of coping strategies.

***Problem-focused coping.*** When fathers view LQTS as a condition that is controllable, they may show a tendency to take an active problem-solving approach to managing this stressor. Problem-focused coping is considered one of the most adaptive styles of coping and may be particularly useful in highly controllable conditions (Riley & Park, 2014). Fathers endorsed problem-focused coping strategies by actively seeking information about LQTS and using that information to make well-informed decisions about many aspects of their children's lives, such as medical treatment and lifestyle choices. The management of LQTS was described by fathers as involving an objective assessment of potential risks posed to their children in various environments and appropriate precautions to avoid or minimize those risks. Rather than absolute avoidance of all activities that could carry a potential risk, fathers preferred a balanced and reasonable assessment of risks and benefits of children's active pursuits.

***Processing emotions.*** Uncertainty is a common component of illness and can be an ongoing source of stress in fathers of children with LQTS. While fathers take active steps to effectively minimize the risks associated with LQTS, cardiac events,

even if unlikely, may be sudden and unexpected and, at worst, fatal when they do occur. Therefore, even when the probability of an event occurring may be perceived as low, fathers' perception of the uncontrollability of a sudden and unexpected event contributes to lingering worries. Furthermore, in a high-stakes situation, such as a child's potentially life-threatening condition, even low-probability events are more likely to be appraised in a threatening manner (Mishel, 1988). According to Mishel (1999), danger appraisals tend to be accompanied by emotion-focused coping strategies, such as avoidance or wishful thinking. While fewer fathers reported direct use of emotion-focused coping methods, a couple of fathers coped with difficult emotions or traumatic memories by trying not to think about distressing emotions or events. Fathers acknowledged their feelings of fear and worry associated with their children's well-being; however, they generally denied the use of or need for particular strategies to process their own emotions. Fathers generally felt capable of managing their own stress and were more concerned with accessing emotional support for their children. While most fathers did not report actively seeking out emotional support from others, they indicated a desire to support others and felt that sharing their story with others was a way in which something positive could emerge from their experiences. While fathers did not report directly focusing and expressing their emotions as a way to cope, they did change their own behaviors by becoming more vigilant in an effort to reduce their fears.

***Meaning-focused coping: A positive reinterpretation of LQTS.*** While LQTS is a highly treatable condition when appropriately diagnosed, fathers do worry about the aspects of LQTS that are perceived as uncontrollable, or the uncertain threat of

symptoms suddenly striking without warning. In addition to problem-focused strategies aimed at actively modifying their environment, fathers also used coping strategies to change their view or perspective. Acceptance- and meaning-focused coping strategies may be useful to fathers as a way to reduce stress by changing their thoughts and beliefs associated with LQTS. Meaning-focused coping includes efforts to cope with difficult emotions by cultivating acceptance and finding meaning and value within the context of stressful life experiences (Roubinov, Turner, & Williams, 2015). Individuals with chronic conditions may demonstrate lower levels of distress when uncontrollable stress is met with predominantly meaning-focused coping (Roubinov et al., 2015). Acceptance as a coping strategy for individuals with a chronic heart condition may diminish psychological distress and increase one's feelings of liveliness and vigor (McCabe & Barnason, 2012). Meaning-focused strategies, such as positive reinterpretation or positive reframing, appear to be especially helpful for situations that are appraised as outside of one's control and may be helpful for managing stress associated with aspects of LQTS that cannot be directly resolved (Linley & Joseph, 2004).

Fathers used meaning-focused strategies by redefining their experiences associated with LQTS. Reframing the discovery of LQTS in a positive manner, fathers viewed themselves as fortunate to have learned of the diagnosis of LQTS, as this discovery provided them with an opportunity to receive proper treatment and avoid potentially tragic consequences. Fathers also developed new perspectives on life with a deeper appreciation for the preciousness of each day and a shifting of priorities based upon a reassessment of values. Positive reinterpretations of LQTS

included fathers' recognition of the ways in which their families had changed for the better, believing that their families had been brought closer together and had become stronger in response to LQTS. Acceptance-based strategies included fathers' ability to adjust their expectations for their children by redefining their priorities, as well as to cultivate an acceptance of the uncertainties of life and events beyond their control.

Religious coping strategies may be helpful in situations of low direct control by providing meaning, hope, and positive perspectives on one's life in the face of adverse circumstances ( Park, 2005). Beliefs about fate may provide emotional strength to families in overcoming challenges believed to be beyond their control. Spiritual or religious coping strategies were demonstrated in 50% of mothers (= 5) whose children were diagnosed with LQTS. These mothers referred to using faith or a strong sense of spirituality or religiosity as a means of coping (Gonzales, 2009). In this particular sample of fathers, only one father described religious coping as a primary method for managing distress.

### **Implications of Research Findings**

While the findings of this study were drawn from a small sample size of fathers, a number of observations can be drawn from qualitative analysis of the data. These findings indicate that fathers are significantly affected by the discovery of LQTS in their children and suggest risk and resistance factors that may influence fathers' adjustment. An understanding of factors that may contribute to heightened distress in fathers of children with LQTS can assist healthcare providers in identifying families at increased risk for adjustment difficulties. Several factors implicated by these findings may contribute to fathers' levels of distress, including the frequency

and severity of children's cardiac symptoms; witnessing their children experience a significant cardiac event, such as cardiac arrest; and ongoing fears associated with the uncertain threat of sudden death. Some fathers may experience the highest levels of distress during the time of their children's diagnosis, when support from healthcare providers may be most beneficial. Knowledge may help to promote a greater level of predictability surrounding a chronic illness and reduce distress by increasing understanding of the feared unknown (Farnsworth et al., 2006; Mishel, 1988; Sterken, 1996).

While rapid advances in the diagnosis and risk stratification of LQTS has helped increase predictability of cardiac events and informed medical treatment, diagnostic and risk assessment is a highly complex process that requires clinical expertise. Fathers are challenged by the complex medical decisions involved in the ongoing treatment and management of LQTS in their children. Fathers may struggle to comprehend the seriousness of LQTS when their children display no symptoms and may question the need for invasive medical procedures. In particular, concerns related to implantation, proper functioning, future replacement, and cost of ICD were frequently cited by fathers. Healthcare providers must be able to translate complex health information to families to facilitate understanding of diagnostic results, associated risks, and recommended treatments and to increase families' capability for informed decision making.

Additionally, fathers said that being provided with the right information at the right times is needed to navigate the many complex aspects of LQTS from the period of diagnosis to ongoing daily management of children and adolescents. Healthcare

professionals should not assume that families understand information that has been provided to them, but should assess families' levels of understanding on a continual basis as families face new challenges and transitions. For instance, fathers may require specific support and guidance when their children are diagnosed with LQTS at birth and require hospitalization and subsequent monitoring at home. Care for infants must be closely coordinated among physicians in NICUs, cardiologists with expertise in arrhythmias or electrophysiologists, and other pediatric healthcare providers. Primary-care physicians working with children should coordinate care with treating cardiologists or electrophysiologists. The needs of adolescents and young adults may also present unique concerns for fathers. The approach of adolescence can be a time of increased stress, as children may be prone to engage in more risky behaviors and spend less time under parental supervision. Fathers worry about older adolescents transitioning to college and being on their own for the first time. Healthcare providers can support fathers during transitional periods by preparing families for these changes and offering recommendations to address their specific concerns.

Fathers desire more for their children than the prevention of symptoms; they want them to enjoy happy, active, and fulfilled lives. Fathers may question whether they are making the right decisions for their children, while healthcare providers can relieve pressure on parents by providing ongoing knowledge and support in the daily management of LQTS. Healthcare providers must support families in a decision-making process that places the highest regard for children's quality of life. Ackerman (2015) proposed that the prevention of sudden death in LQTS has become readily

attainable and that individualized treatment for LQTS should enable children to live and thrive despite their diagnosis. Ongoing knowledge and support will assist fathers seeking to achieve the best quality of life for their children by helping them to achieve a healthy level of caution through informed decisions.

Historically, child psychology and pediatric psychology research studies have relied upon mothers' perspectives while leaving out the experiences of fathers who have children with chronic health conditions. Most fathers in this study were highly involved in children's activities and pursuits and viewed themselves as working with their spouses as a team in making decisions and managing responsibilities associated with their children's LQTS. While fathers want to be informed and involved in their children's overall treatment, some may regard themselves as outsiders. Fathers may not attend children's medical appointments as frequently as mothers and therefore may rely upon their wives for information. Healthcare providers may best address the needs of all family members by assessing all family members' level of understanding and providing alternative methods of communicating other than directly through medical appointments. In fact, many fathers expressed strong and mostly positive feelings about their children's physicians and appreciated their accessibility through e-mail. Regardless of whether or not these fathers represent the majority, there are fathers who wish for a higher level of contact and involvement.

Another point of inquiry was fathers' coping styles. Previous studies in the pediatric literature suggest that fathers may use a more practical or instrumental style of coping, whereas mothers are more attuned to affective issues (Barak-Levy & Atzaba-Poria, 2013; Hoekstra-Weebers, Jaspers, Kamps, & Klip, 1998). The

overall findings of this study suggest, however, that fathers have a number of both problem-focused and emotion-focused strategies available to them and use a range of coping strategies. Fathers may prefer a subset of emotion-based coping strategies that emphasizes the use of cognitive coping strategies or positive reframing strategies rather than strategies involving the processing and expression of emotions. Previous research investigating coping styles suggests that different strategies may be useful, depending on the situation and context and the degree to which stressors are perceived as controllable (Riley & Park, 2014). Fathers may benefit from flexible use of coping styles in managing stress associated with LQTS that involve both behavioral or active problem-focused strategies and cognitive or internal coping processes to alleviate psychosocial stress. Healthcare providers may benefit from tailoring interventions for fathers by assessing their particular concerns regarding their children and identifying appropriate coping responses. As noted, fathers may experience uncertainty and associated stress related to various aspects of LQTS, such as the seriousness of the condition, the risk of cardiac events, the ambiguity of symptoms, the necessity for treatments, and the implications of the condition on their children's futures. Providing knowledge and information based upon fathers' specific areas of concern may help reduce uncertainty, while fathers may benefit from coping strategies designed to facilitate adaptation over the trajectory of their children's condition by learning to accept and manage uncertainty itself (Kerr & Haas, 2014).

## **Implications of Research Findings as Related to Diversity**

Fathers' coping and adaptation to their children's chronic health condition occur within a context of cultural and ethnic background through which one learns responses to health and illness (Desai, Rivera, & Backes, 2015). The current study included a small sample of a mostly culturally and ethnically homogeneous group of individuals, while the ethnic background of fathers participating in the online survey was unknown. All of the fathers interviewed were Caucasian and living in the United States, with the exception of one father who was of greater Middle Eastern descent and living in his native country. While fathers' educational backgrounds ranged from high school to college, most fathers living in the United States were earning in a middle-class to upper-middle-class range. Because of this small and limited sample size as related to ethnic and cultural factors, the findings of this study in relation to issues of diversity are also limited. These findings did reveal, however, a striking difference between the medical resources available to individuals known to be living within the United States and resources available to the single participant known to be living in a greater Middle Eastern country. This difference underscored disparities in the availability and accessibility of healthcare between countries and the significant impact this reality has on families of children with serious health conditions. Interestingly, access to the Internet proved critical in obtaining medical equipment and the medical expertise of physicians within the United States to an extremely resourceful father living outside of the United States. Whether or not disparities exist within the United States in the accessibility to advanced medical care

among families of diverse cultural and ethnic backgrounds affected by LQTS is an issue that needs further investigation.

### **Implications of Research Findings as Related to Advocacy**

LQTS is a potentially lethal cardiac condition that may be responsible for as many as 3,000 unexpected deaths in children and young adults in the United States per year (Modell & Lehmann, 2006). Rapid advances in the detection and risk stratification of this cardiac arrhythmia syndrome have resulted in highly effective treatments. Despite this tremendous progress in the understanding and management of this cardiac condition in the past 2 decades, LQTS remains relatively unknown to the general population. Nearly all of the fathers in this study had no knowledge of LQTS prior to their children's diagnosis. While it remains a less widely known condition, a significant portion of sudden unexplained deaths may be secondary to such primary arrhythmia syndromes as LQTS. Fathers feel strongly in favor of the delivery to families of preventative and timely education regarding LQTS to avoid the potential devastation of an unexpected death in an otherwise healthy young individual. Several fathers described their wish to share their stories with others so that other families could benefit from what they had learned through their children's experiences. In fact, fathers who were interviewed unanimously hoped that their participation in this study would facilitate greater awareness and understanding of LQTS within and beyond their own communities. Fathers were supportive of LQTS groups dedicated to raising awareness of LQTS among medical providers and individuals and that support initiatives, such as cardiac screenings for young children and athletes. The critical support that online groups can provide to families with

limited access to advanced healthcare was stressed by one father living outside of the United States who believed that, without access to the Internet, Facebook, crowd-funding sources, and other online LQTS groups, he could not have attained a proper diagnosis and received the necessary treatment for his daughter.

### **Limitations of the Current Study**

While the research findings revealed important trends in fathers' responses and experiences related to their children's LQTS, the study does present some limitations. The sample size of this study was small and predominantly Caucasian among participants whose race was known. Most participants reported incomes of \$75,000 or above. All but one father interviewed lived within a limited geographic area of the United States. Owing to the homogeneous nature of the study sample, the findings may not be generalizable and fully represent the population of interest. Specifically with regard to the ethnic and racial breakdown of the sample size, the findings may be limited regarding paternal roles that may be specific to a particular culture, but may not be true for other cultures or one's ethnicity. Furthermore, the participants in this study were comprised of a self-selected sample of fathers who may have responded to research notices on LQTS websites or social media sites requesting volunteers and are a representation of those fathers actively seeking information and willing to participate. Fathers who have more difficulty adjusting to their children's illness or hold more negative perceptions might have been less likely to participate in the study (Knafl & Zoeller, 2000). Another possibility, as suggested by the literature, is that fathers have varying degrees of flexibility in their work settings, thus impacting their ability to participate in research

studies. Because fathers with higher paying jobs and with higher social status may be afforded greater flexibility, low socioeconomic status emerges as a risk factor for limited involvement (McNeill, 2004). Furthermore, few participants in this study reported low income, while individuals in lower socioeconomic strata may be vulnerable to higher levels of distress caused by adverse life events, lower perception of control, and reduced cognitive flexibility to cope with stressors (Hendriks et al., 2005; Oosterwijk, 2004; Schein, Bernard, Spitz, & Muskin, 2003).

### **Suggestions for Future Work**

The present study was an initial exploration into fathers' experiences, concerns, and ways of coping with their children's LQTS. While the sample of participants was small, the findings from this investigation revealed possible critical factors relating to the psychological adjustment of fathers that deserve the attention of future research. A larger, diverse, and more representative sample of the population of interest will assist in further investigation and elaboration of factors influencing fathers' psychosocial adjustment to LQTS. Additional research may help elucidate critical factors that may impact fathers' adaptational processes, including LQTS-related factors, such as frequency and severity of children's symptoms; treatment factors; lifestyle factors and restrictions; and fathers' cognitive appraisals and coping styles.

Additional research should investigate relationships between fathers' coping and adjustment and children's psychosocial adjustment. As noted, previous studies suggest that fathers' psychosocial adjustment had a significant influence on children's adjustment to chronic health conditions and that children may be more sensitive to

fathers' distress as compared to mothers' distress. Currently, no studies have investigated the relationship of fathers' coping and the adjustment of their children with LQTS, while further research in this area could improve psychosocial outcomes for children with LQTS and their families.

Future studies should investigate the effectiveness of interventions provided by medical and healthcare professionals that are tailored to fathers' needs. Research in this area should include investigation of outcomes associated with education-based interventions aimed at counseling families and fathers within a patient/family-centered decision-making model. Clinical studies should also investigate the effectiveness of cognitive interventions aimed at reducing fathers' stress through cognitive reframing techniques and assisting fathers in redefining their experiences associated with LQTS. Acceptance and commitment therapy interventions that facilitate acceptance of distressing thoughts and emotions while engaging in value-driven behaviors may be of particular use for individuals faced with uncertain situations, such as chronic illness, and studies investigating use of such strategies in coping with LQTS may be warranted (Hayes, Strosahl, & Wilson, 1999; Burke et al., 2014).

### **Conclusion**

This study was designed to examine the unique experiences of fathers of children with LQTS and used a qualitative study to better investigate the phenomenon of interest. The results from this study revealed a more in depth understanding of the nature of fathers' concerns, suggested interventions tailored to fathers' needs, and may promote further research. The fear and uncertainty that families face when their

children are diagnosed with LQTS can be overwhelming. The management of symptoms through lifestyle changes and medical treatment can involve complex decisions that are challenging for families to navigate. Fathers strive to seek a balance in protecting their children from harm while maintaining as normal a lifestyle as possible in which their children are happy and thrive. Concerns about their children revolve around both the short-term and long-term implications of LQTS, and fathers worry about their children's future. Recognizing fathers' interest in gathering information and problem solving may encourage healthcare providers to seek out the involvement of fathers and may involve identifying modes of communication best adapted to each family. Increased communication could benefit fathers by including them in the decision-making process, leading to a positive impact on the entire family and strengthening the capacity of the family to adjust effectively. Furthermore, fathers play a distinct and important role in their children's lives. The emotional well-being of fathers needs attention, as their positive involvement influences their children's social, emotional, and cognitive development. By attending to the particular needs of fathers, healthcare providers can increase the likelihood that fathers remain involved with their children in a positive manner.

While many fathers experience increased levels of distress related to their children's chronic health illness, these findings also suggest that many fathers adjust effectively and engage in healthy coping strategies. The results of this study may provide information regarding the particular approaches that well-adjusted fathers use in managing and coping with their reactions to LQTS. These findings could be

helpful in developing interventions designed to promote the positive adjustment of both fathers and mothers living with LQTS in their families.

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APPENDIX A  
Web-based Questionnaire

1. When did you first become aware of LQTS in your family?
2. Describe your initial reaction when your child was diagnosed with LQTS.
3. What has been the most difficult aspect of the diagnosis of LQTS?
4. Describe your greatest concerns regarding your child and his/her condition.
5. Has LQTS restricted your child from extra-curricular or physical activities?
6. Describe resources which have been the most helpful to you and your family.
7. How has your experience been with your child's health care team?
8. If there is anything you would like to comment on further or share about your experiences, please do so here.

## APPENDIX B INVITATION TO FATHERS

Dear Fathers,

I am writing to tell you about a study aimed at better understanding the experiences of fathers of children with Long QT Syndrome (LQTS). This study will specifically examine the concerns of fathers of children with LQTS, and the challenges they may encounter. You may be eligible to participate in this study. You are being contacted because your child or children have been identified as having LQTS and therefore you will be able to take part in this research. We are contacting fathers that have a child who has been diagnosed with LQTS for at least one year. Fathers who choose to participate in this study will be interviewed by a doctoral candidate in psychology attending the Philadelphia College of Osteopathic Medicine (PCOM). Each interview will consist of several questions to learn about the experiences fathers have had with LQTS and how it has affected family life. The questions are "open-ended," which means that they invite more than a "yes" or "no" answer. When the interview is finished, a brief questionnaire will be completed which will take about 10-20 minutes to complete. While each interview will be audio-recorded, no names will be associated with the recorded material. Fathers may not gain any direct benefit from participating, however, the final results may help to identify the psychological and social needs of fathers affected by LQTS in their families, and may benefit families in the future. No individual feedback will be given to participants, or to their doctors.

If you want to learn more about this project, please send an email to the research coordinator at [LQTstudies@pcom.edu](mailto:LQTstudies@pcom.edu) and someone will contact you promptly to describe the study in detail. Alternately, you may leave a voice mail at (215) 871-6464 and someone will return your call promptly. Any questions you may have will be answered at this time. If you do not respond with an email or phone call, you will not be contacted again. We hope that you are interested and will participate in this important research. You may call the number listed above at any time for more information. Thank you for your time and interest.

Sincerely,

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Victoria L. Vetter, M.D., MPH

Director Youth Heart Watch, Children's Hospital of Philadelphia

## APPENDIX C

### INTERNET RECRUITMENT ANNOUNCEMENTS: INTERVIEWS

#### **Twitter - LQTstudies.com Twitter account**

LQTstudies Group at the Philadelphia College of Osteopathic Medicine, researching the experiences of fathers 18+ who have children with Long QT Syndrome. Email [lqtstudies@pcom.edu](mailto:lqtstudies@pcom.edu) Can you help with a RT please?

#### **LQTS-related Websites**

##### **Fathers of Children with Long QT Syndrome**

**FATHERS of children with Long QT Syndrome** are needed to share their experiences. To participate in this research study, fathers must be over the age of 18 with at least one child who has been diagnosed with LQTS syndrome for at least 1 year.

Participation involves a 60-90 minute interview in person or via telephone or video call. The purpose of this study is to examine how families are affected by LQTS by describing the perspectives of fathers. You will be asked several questions about the challenges that you have encountered as a parent of a child with LQTS and how you have coped with these challenges. You will also be asked to complete a brief demographic questionnaire.

Participants will receive two movie tickets when the interview is completed.

##### **Contact Information**

The principal investigator of this study is Stephanie Felgoise, Ph.D., ABPP at the Philadelphia College of Osteopathic Medicine (PCOM). To ask questions about the study, please email Jodi Yarnell, Ed.S. at [lqtstudies@pcom.edu](mailto:lqtstudies@pcom.edu) or call (215) 871-6464 and leave a message. Someone will contact you promptly.

## APPENDIX D

### INTERNET RECRUITMENT ANNOUNCEMENT: WEB-BASED QUESTIONNAIRE

#### **Fathers of Children with Long QT Syndrome**

**FATHERS of children with Long QT Syndrome** are needed to share their experiences. To participate in this research study, fathers must be over the age of 18 with at least one child who has been diagnosed with LQTS syndrome for at least 1 year.

Participation involves an online questionnaire consisting of open-ended questions related to LQTS in your family. The purpose of this study is to examine how families are affected by LQTS by describing the experiences of fathers who have children diagnosed with this condition. Study information is ANONYMOUS and any information obtained through the online questionnaire cannot be linked to you in any way. If you would like to participate in the study, click the link below: <https://www.surveymonkey.com/s/LGNQCF3>

#### **Contact Information**

The principal investigator of this study is Stephanie Felgoise, Ph.D., ABPP at the Philadelphia College of Osteopathic Medicine (PCOM). To ask questions about the study, please email Jodi Yarnell, Ed.S. at [lqtstudies@pcom.edu](mailto:lqtstudies@pcom.edu) or call (215) 871-6464 and leave a message. Someone will contact you promptly.

## APPENDIX E

### PERSONAL INFORMATION QUESTIONNAIRE

FATHERS OF CHILDREN WITH LONG QT SYNDROME:

A QUALITATIVE STUDY OF THEIR EXPERIENCES

**Study ID:**

**Date:**

#### PERSONAL INFORMATION REGARDING YOU

Your Age \_\_\_\_\_

Ethnic Background:

Asian  African American  Caucasian  Indian  Latino  Other

Estimated Household 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 currently working?  Yes If so, how many hours? \_\_\_\_\_  No

Do you carry the diagnosis of LQTS?  Yes  No

If so, are you symptomatic?  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 1 year and 17 years of age?  
\_\_\_\_\_

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

At what age did your 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.

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

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Briefly describe the symptoms:

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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?

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 has your child/children had?

never 1-3     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 your child/children has a pacemaker or implanted cardioverter defibrillator (ICD), does it create any difficult issues or problems for you? Please briefly explain.

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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)

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

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Has genetic testing been done? yes no    If so, who else has LQTS in your family? \_\_\_\_\_

THANK YOU VERY MUCH FOR YOUR PARTICIPATION!