

2010

School Accommodations to Support Psychosocial Functioning and Self-Esteem in Adolescents with Long QT Syndrome

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Philadelphia College of Osteopathic Medicine

Department of Psychology

SCHOOL ACCOMMODATIONS TO SUPPORT PSYCHOSOCIAL FUNCTIONING
AND SELF-ESTEEM IN ADOLESCENTS
WITH LONG QT SYNDROME

By Betsy E. Feinberg, M.S.Ed., M.S.

Submitted in Partial Fulfillment of the Requirements of the Degree of

Doctor of Psychology

July 2010

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

Dissertation Approval

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

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ACKNOWLEDGEMENTS

I would like to thank everyone who supported me and guided me through my dissertation process. First, Dr. Felgoise, who was a tremendous mentor and guide throughout my graduate years. I aspire to have the qualities she possesses, including drive, vision, perseverance, and dedication to her students and research. I admire her flexibility, thoughtfulness, and integrity. I am grateful to have had the opportunity to expand my interests and experiences with the inspiring population. Secondly, my two committee members, Drs. Salzer and Vetter, who have been flexible and supportive of my dissertation research through every step of the process. I thank them for opening my eyes to the medical and developmental perspectives of adolescents with LQTS.

I must thank my husband, Richard Feinberg, who lovingly shared the computer for years and always encouraged me to achieve my goals. Rich was my greatest source of strength and optimism, always reinforcing my abilities and drive. I must recognize my parents, Sheila and Rick Sapphire, who wondered if there was anything left to learn, but always reminded me of how proud they were and encouraged me to seek my dreams. Last but not least, I must thank my grandma, Trudy Manashil, who has always supported and believed in me, no matter how big or small the task. I am grateful and humbled to have such supportive and caring mentors, teachers, and family.

ABSTRACT

The current manuscript, as part of a larger program of research, intended to examine whether long QT syndrome (LQTS), a chronic and life-threatening genetic cardiac illness, impacts adolescents' psychosocial functioning. After 2 ½ years of involvement in the study and 13 months of attempted data collection, it was determined by the research team that the benefits of proceeding with an intervention plan outweighed the costs of waiting for data. Although the study was not able to be conducted within the anticipated time frame due to unforeseen logistical challenges, the study hypothesized that adolescents with higher ratings of self-reported social and school functioning have higher levels of self esteem (SE) and that social and co curricular restrictions, school accommodations, age of diagnosis, medical event frequency, social functioning, and school functioning predict ratings of SE. Given the hypothesized needs of the population and support based on previous literature and corroborating theories, this manuscript resulted in a detailed school-based accommodation plan, along with a crisis response plan, which will ultimately help advocate for the needs of adolescents with LQTS. The intervention plan will be revised as planned future work, once data has been obtained and analyzed and/or the intervention plan has been field tested.

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EPIGRAPH

“We don’t see things as they are, we see them as we are.” (Anaïs Nin)

PSYCHOSOCIAL FUNCTIONING

CHAPTER 1

Introduction

Healthy adolescence is a time of flourishing development, involving social and emotional milestones, such as establishing same-sex and opposite-sex friendships, attending school, participating in social activities, and becoming active in co curricular activities, such as sports and art (Vitulano, 2003). In addition to the unique developmental and maturational challenges that adolescents face, adolescent chronic illness (CI) can pose stumbling blocks for youth. For instance, adolescent CI has been associated with adjustment problems and internalizing disorders, such as anxiety, depression, and eating disorders (Vitulano). CI in adolescence has also been linked with behavioral disorders, psychiatric diagnosis, and somatic symptoms (Suris, Michaud, & Viner, 2004). Since certain medical conditions can hinder participation in social and school activities, it is critical to discover which diagnoses pose challenges for adolescents and target those individuals for primary or secondary interventions to help aid in the successful navigation of this unique developmental stage.

Statement of the Problem

Long QT Syndrome

About the condition. Long QT syndrome (LQTS) is an inherited genetic cardiac syndrome, characterized by prolonged intervals between the discharging and recharging of heartbeats, otherwise known as QTc interval prolongations (Arnestad, Crotti, Rognum, Insolia, Pedrazzini, Ferrandi et al., 2007; Bonn, 1997). This electrical cardiac instability places LQTS patients at increased for irregular heart rhythms and sudden cardiac arrest,

although they do not have structural abnormalities of their hearts (Napolitano, Bloise, & Priori, 2006). The deleterious effects of LQTS can be triggered by physical exertion, such as swimming, sudden auditory stimuli (loud noises from alarm clocks, telephones, or doorbells), emotional stress, fright, anger, and sleep (Collins & Van Hare, 2006; Kapetanopoulos, Kluger, Maron, & Thompson, 2006). For instance, the condition has been linked to sudden death in young athletes due to participation in competitive and rigorous athletics (Corrado, 2005). Hence, individuals with LQTS are often restricted from engaging in activities that could lead to sympathetic nervous system activation and heart rate acceleration (Corrado). Therefore, LQTS limitations may present obstacles to attaining adolescent milestones, such as establishing friendships, engaging in athletics, or school functioning. Researchers have found that physical self-esteem (SE) mediates general SE (Bowker, 2006), and low levels of academic achievement are associated with lower SE among Portuguese adolescents (Alves-Martins, Peixoto, Gouveia-Pereira, Amaral, & Pedro, 2002). Low ratings of SE have also been linked with stressful life events, poor identity in social groups, unattained personal goals, depression, and anxiety (Cast & Burke, 2002; Youngs, Rathge, Mullis, & Mullis, 1990). Since patients with LQTS may consider their condition stressful, might have limited social groups due to social and athletic restrictions, and often have restrictions from participating in co curricular and athletic activities, adolescents with LQTS may be at risk for reduced SE, making them a potentially vulnerable population, warranting school-based services and crisis plans.

Research needed. Currently, there is minimal research investigating the social functioning, school functioning, or SE of adolescents with heart disease. Furthermore,

this is one of the first manuscripts pertaining to the psychosocial functioning of individuals with LQTS. Given the tenuous development of adolescents, coupled with the social and physical restrictions that adolescents with LQTS face, this population could benefit tremendously from support from the psychological and school communities.

Purpose of the Study

The purpose of the current pilot investigation was to examine whether the social restrictions and psychological characteristics imposed by LQTS predicts adolescents' self-esteem (SE) and to stimulate the creation of school-based accommodations and encourage advocacy for adolescents with LQTS. Although the study was not able to be conducted due to unforeseen logistical challenges within the anticipated timeframe, it was hypothesized that the unique challenges that LQTS imposes on adolescents may impact their social functioning, school functioning, and SE. Specifically, it was hypothesized that adolescents with higher ratings of self-reported social and school functioning would have higher levels of SE. Additionally, it was hypothesized that social and co curricular restrictions, school accommodations, age of diagnosis, frequency of medical events, social functioning, and school functioning contribute to the prediction of SE. Although data was not collected, literature pertaining to adolescent chronic illness and LQTS studies supports the need for interventions and school-based accommodation plans. After 13 months of attempting to establish data collection venues as part of a larger program of research, it was determined that the benefits of proceeding with an intervention plan outweighed the costs of waiting for data for the purposes of the population and of this

manuscript. The original study and the larger program of research are currently under way.

Ultimately, the goal of this manual is to provide schools, medical professionals, and families guidance in creating and implementing comprehensive school-based accommodation plans and advocating for the needs of adolescents with LQTS. It is the hope that recommendations from this manuscript may lead to increased education, research in the areas of needs assessment, school-based protocols, and future studies. Accommodation plans from this thesis may sensitize practitioners to the implications of living with LQTS.

Overview of Literature Review

First, this manuscript will review the characteristics and symptoms of LQTS and the challenges that patients face, including treatments, restrictions, the unpredictability of the syndrome, and medical adherence. Secondly, theories of healthy adolescent self-esteem, including SE constructs, social perception theories, social comparison theory, and cognitive theory, will be examined. There will also be a discussion about the potential impact of LQTS on adolescent SE, including social restrictions, activity restrictions, school accommodations, age of diagnosis, and frequency of events. Existing literature pertaining to adolescent chronic illness (CI) and SE will be explored. Lastly, a comprehensive school-based accommodation tool and emergency response plan will be introduced.

CHAPTER 2

Literature Review

*Rationale for Target Population**LQTS Characteristics*

LQTS definition. An inherited genetic cardiac condition, long QT syndrome (LQTS) is characterized by prolonged intervals between the discharging and recharging of heartbeats, otherwise known as QTc interval prolongations (Arnestad et al., 2007; Bonn, 1997). This electrical instability is caused by cardiac gene ion channel mutations, and can lead to sudden death from aborted sudden cardiac arrest or malignant ventricular tachyarrhythmia (rapid heartbeat), including torsade de pointes, which is a transient malignant arrhythmia characterized by irregular heart wave amplitude changes (Bonn; Moss, Zareba, & Robinson, 1995; Vetter, 2007; Viskin, 1999). This electrophysiological deficit also predisposes individuals to syncope, which is loss of consciousness (Collins & Van Hare, 2006; Garson, Dick, Fournier, Gillerre, Hamilton, Kugler et al., 1993). Although LQTS patients are at increased risk for life-threatening arrhythmias, which are irregular heart rhythms, they do not have structural abnormalities of their hearts (Napolitano, Bloise, & Priori, 2006).

Symptoms and manifestations. The symptoms of LQTS include pathological heart repolarization and prolonged QTc intervals (Hobbs, Peterson, Moss, McNitt, Zareba, Goldenberg et al., 2006). The three hallmarks of LQTS are QTc prolongation, unexplained fainting, or premature sudden death in patients or family members of patients (Moss et al., 1995). Children and adolescents often present with syncope during or immediately following exercise, such as swimming, or physical or emotional stress

(Vetter, 2007; Viskin, 1999). Arrhythmias also tend to occur after arousal from noise and during sleep (Viskin). Garson et al. (1993) found that of initial presentations among children, 67% included symptoms from exercise, 39% had a family history of prolonged QTc, and 31% had a family history of sudden death. Additionally, LQTS patients may initially present with ventricular tachycardia or an aborted sudden cardiac arrest (Vetter).

LQTS patients are diverse, as their clinical courses often differ, even among family members in whom it has been diagnosed (Moss et al., 1995). For instance, symptomatic patients tend to present with unexplained fainting spells, palpitations, seizures, or cardiac arrest (Hobbs et al., 2006). For others, their rhythm disturbances do not cause symptoms, making it less likely their condition will be suspected (Collins & Van Hare, 2006). For instance, Vincent (2003) stated that approximately 50% of LQTS carriers never have symptoms. Unfortunately, sudden death may be the first sign of the syndrome for these individuals, as 14% of all patients die during their first cardiac episode (Collins & Van Hare; Schwartz, Priori, Dumaine, & Napolitano, 2000). Corrado, Basso, and Thiene (2005) concurred that sudden death during athletic activity is often the first manifestation of cardiovascular illness.

Due to the existence of the condition, irrespective of symptom presentation, Garson et al. (1993) stated that children with a prolonged QTc interval and a positive family history of LQTS should be treated, regardless of symptoms. These researchers found that 12% of asymptomatic patients with a prolonged QTc interval and a positive LQTS family history later developed symptoms, and 4% had sudden death (Garson et al.).

Prevalence. It is estimated that LQTS affects approximately 1 in 2,500 people and is a primary cause of sudden cardiac death in youth (Berul, 2008; Collins & Van Hare, 2006). It has also been estimated that an additional 3,000 unexplained deaths each year in the United States should be attributed to LQTS, but the condition is misdiagnosed or undiagnosed (*Long QT Syndrome*, 2008). In their study, Schwartz, Priori, and Napolitano (2003) found that the LQTS prevalence rates are likely much higher than researchers currently hypothesize, and a relatively high number of “healthy” individuals have an underlying susceptibility to electrical cardiac instability, but do not have prolonged QT intervals. Hence, the exact prevalence rates of the syndrome are still not known because the literature lacks large-scale prospective population studies, although it is believed that the syndrome is more common than what current statistics indicate (Kapetanopoulos et al., 2006; Schwartz et al., 2003).

Etiology. LQTS is a heritable condition that is caused by genetic mutations in the potassium-channel genes (Moss et al., 1995). Although the inheritance in the majority of the cases is autosomal dominant, each child of an affected parent having a 50% chance of inheriting the gene, it can also be recessive, with a 25% chance of inheriting the gene (Imboden, Swan, Denjoy, & Van Langen, 2006). Furthermore, sporadic nonfamilial cases appear in approximately 10% of the reported LQTS population, suggesting non inherited genetic mutations (Moss et al.). QTc prolongation may also be secondary to certain medications, such as antibiotics, antihistamines, serotonin inhibitors, or tricyclic antidepressants, although an underlying susceptibility to arrhythmias or torsade de points often exists (Viskin, 1999). Furthermore, LQTS can be acquired through metabolic

disorders, electrolyte abnormalities, altered nutritional states, poisons, or intracranial diseases or injuries (Moss et al., 1995; Napolitano et al., 2006).

Variations. Historically, when first described in the late 1950s and early 1960s, LQTS was thought to be extremely rare and consist of two primary designations based on inheritance type and the presence of deafness, which were the Romano-Ward syndrome and the Jervell and Lange-Nielson syndrome (Collins & Van Hare, 2006; Kapetanopolous et al., 2006). Current researchers have expanded these initial findings, discovering that LQTS is a genetically heterogeneous syndrome, consisting of at least 500 mutations in 10 different genes, all containing clinical variability (Patel & Antzelevitch, 2008). These genetic variations are named LQT1 through LQT12 (Zareba & Cygankiewicz, 2008). LQTS1 is most common, followed by LQTS2 through LQTS7, which is a rare condition (Kapetanopolous et al.). Although all LQTS variants are characterized by prolonged ventricular repolarization, they are associated with varying triggers and risk factors (Kapetanopolous et al.).

Triggers. The different LQTS genotypes, or genetic variations, are associated with varying phenotypes, or clinical presentations. Triggers of cardiac events often correlate with one's genetic LQTS variant (Kapetanopolous et al.). For instance, LQT1 is associated with sudden death from exercise, such as swimming, individuals with LQT2 are more likely to have events caused by sudden auditory stimuli, such as a loud noise or emotional stress, and LQT3 is associated with sudden death during sleep (Collins & Van Hare, 2006; Vetter, 2007). Conversely, the literature also makes note of genetic overlaps of these variations and the imprecise association between genotype and clinical presentations (Collins & Van Hare). Therefore, although distinct genetic mutations have

been discovered, they are not mutually exclusive and contain both clinical variability and similarity.

Risk stratification. There are numerous LQTS risk factors, including family history, age, gender, genotype, and phenotype. Risk stratification is often based on timing and frequency of recent syncope, the duration of the QTc interval, and sex (Hobbs et al., 2006). For instance, LQTS patients who develop syncope or aborted cardiac arrest, especially during childhood and adolescence, have a 4% to 6% likelihood that their symptoms will recur that year (Moss et al., 1995). Others have found a positive correlation among likelihood of cardiac arrest and length of QTc interval prolongation (Priori, Schwartz, Napolitano, Bloise, Ronchetti et al., 2003). Garson et al. (1993) concluded that children with extremely long QTc intervals, greater or equal to 500 milliseconds, are at high risk for sudden death. Imboden et al. (2006) found a female predominance in symptomatic LQTS patients. This was hypothesized to occur due to a longer QTc interval in women, the expression of potassium-channel genes in females, a predisposition to torsade de pointes in females, and the reduced effectiveness of beta-blocker treatments in women (Imboden et al.). Priori et al. stated that female gender was associated with a higher incidence of cardiac arrest or sudden death with LQT2, and male gender carried a higher risk in LQT3. Locati et al. (1998) also found that the risk for cardiac events was higher in males until puberty and higher for females in adulthood. In their study, Goldenberg et al. (2008) found that QTc interval prolongation and prior syncope were risk factors for preadolescent boys, while prior syncope was the only risk factor for preadolescent girls. Although the mechanisms are not fully understood, researchers concluded that the rate of fatal or near-fatal cardiac events is higher for boys

throughout childhood, but becomes higher for females throughout adolescence and adulthood (Goldenberg et al.). It was hypothesized that environmental factors, genetic factors, hormones, and levels of physical activity contribute to these age and gender-related risk factors (Goldenberg et al.).

Looking at family history, first degree relatives of a family member who died from LQTS are at high risk for developing life-threatening tachycardia (Moss et al., 1991). Overall, the highest-risk group is LQTS patients who present with syncope do not receive a diagnosis or treatment. Within this subset, up to 20% of patients will have cardiac death in the first year after syncope, and 50% will have sudden cardiac death within 5 years (Vetter, 2007). Priori et al. (2003) stated that these risk stratifications should be evaluated when determining how to treat patients, as proper diagnosis and treatment is critical for these patients.

Diagnosis. Diagnostic criteria for LQTS include a prolonged QTc interval on an electrocardiogram (ECG), syncope, family history of LQTS, or abnormal T waves (Garson et al., 1993). LQTS is often diagnosed through an evaluation of an ECG, based on a corrected measurement of the QTc interval (Arnestad et al., 2007; Napolitano et al., 2006). Individuals who experience syncope during or after exercise, in relation to stress, or who had an aborted cardiac arrest should undergo an ECG (Vetter, 2007). It has been suggested that athletes with a family history of sudden cardiac death, cardiovascular disease, personal history of unexplained syncope, or resuscitated cardiac arrest should undergo an ECG workup (Kapetanopolous et al., 2006). Vetter also recommended that all family members of those with LQTS should be evaluated.

A significant number of LQTS carriers do not demonstrate symptoms or a QTc prolongation on ECGs, meaning the gene has not fully expressed itself, although the syndrome is present (Kapetanopolous et al., 2006). Therefore, family histories of unexplained syncope, early sudden cardiac death, or arrhythmias are factors to consider when making a diagnosis (Kapetanopolous et al.). For instance, Vetter (2007) stated that familial questions pertaining to unexplained fainting, seizures, deafness, sudden infant death syndrome, unexpected deaths, deaths from car accidents or swimming, and LQTS history can provide critical diagnostic information. Since LQTS is a genetic condition, ECGs are often conducted on family members to assist in making a diagnosis (Kapetanopolous et al.). Interestingly, approximately 60% of patients with LQTS are identified after LQTS has been diagnosed in a family member (Moss et al., 1995). For instance, another study found that the most common reason for diagnosis was an abnormal ECG during a family screening, which has lead to earlier identification and treatment (Petko et al., 2008). Hence, LQTS is often diagnosed in asymptomatic carriers by ECG testing for unrelated reasons or through family member screenings (Kapetanopolous et al.).

In recent years, genetic testing has become available, making LTQS one of the first cardiovascular diseases to utilize molecular testing (Collins & Van Hare, 2006). Although this testing has allowed for accurate diagnoses and treatment recommendations, especially for borderline or asymptomatic individuals, a negative test result does not necessarily indicate the absence of the condition, since undiscovered gene variations may exist (Collins & Van Hare). Vetter (2007) stated that 30% of genes or gene locations have yet to be discovered. Additionally, Collins and Van Hare stated that genetic tests can only

detect LQTS in approximately 75% of families. Therefore, for 25% of families, this test will not provide definitive results. Furthermore, clinical applicability is reduced, due to the high cost of genetic testing and limited availability (Kapetanopolous et al., 2006; Vetter).

In making a diagnosis, doctors may utilize additional assessments, especially if genetic testing is unattainable. For instance, a 24-hour ambulatory Holter monitor or an exercise stress test may identify arrhythmias, rapid palpitations, or syncope that was not indicated on an ECG (Garson et al., 1993). Post mortem, molecular autopsies are also available to detect LQTS, after which first-degree relatives often go through a detailed assessment (Lee & Ackerman, 2003). Once a diagnosis is made, it is imperative that patients receive proper treatment and recommendations to reduce their cardiac risks.

Treatments. There are currently three accepted therapies for inherited LQTS, including beta-blockers, the implantation of an internal cardioverter-defibrillator (ICD), and restrictions from competitive athletics (Daubert, Zareba, Rosero, Budzikowski, Robinson et al., 2007; Kapetanopolous et al., 2006). Beta-blockers are a class of drugs that slow the heart rate to help prevent tachycardia and sudden cardiac arrest (*Long QT Syndrome*, 2008). Beta-blocker therapy is often utilized as the first line of therapy for LQTS patients, especially when the risks of surgery are too high (Daubert et al.; Kapetanopolous et al.). Beta-blocker therapy, at the maximum tolerated dose, is often the recommended treatment, especially for patients with LQT1 (Patel & Antzelevitch, 2008; Viskin, 1999). Alternately, Patel and Antzelevitch (2008) stated that Beta-blocker therapy should be used with caution in LQT3 patients due to differences in heart repolarization. In their study, Schwartz, Zaza, Locati, and Moss (1991) found that the mortality rate of

those treated with Beta-blockers was 6%, compared to 70% in the untreated population. Hobbs et al. (2006) also found a 64% reduction in life-threatening events in adolescents with LQTS taking Beta-blockers. Although very effective, this treatment does not protect all LQTS groups from cardiac arrest or sudden cardiac death (Kapetanopolous et al.). Therefore, additional treatment options can be added or used as alternatives to Beta-blocker therapy (Kapetanopolous et al.). In their retrospective study, Vincent, Schwartz, Denjoy, Swan, Bithell, Spazzolini et al. (2009) found the reasons for Beta-blocker failure among a database of 216 patients with LQT1 were noncompliance with the Beta-blocker regimen and concurrent administration of drugs known to prolong QTc intervals. Hence, before considering alternate treatments, medical professionals should consider medication compliance, the use of QTc-prolonging drugs, and educating patients on the importance of medical adherence (Vincent et al.).

Implantable cardioverter defibrillators (ICDs) are cardiac devices which are implanted under the skin in the chest to monitor and stop potentially fatal arrhythmias (*Long QT Syndrome*, 2008). They are often utilized after a cardiac arrest, if patients experience recurrent syncope on Beta-blocker therapy, or for high-risk individuals (Collins & Van Hare, 2006; Kapetanopolous et al., 2006). This option should be considered when Beta-blocker treatment is ineffective, since unsuccessful medicine trials are predictors of symptoms and sudden death (Garson et al., 1993). For this reason, in these circumstances, considerations for nonpharmacological treatments, such as a pacemaker or an ICD, are potentially life-saving alternatives (Garson et al.). Alternately, Viskin and Halkin (2009) suggest that studies continue to investigate whether ineffective treatments are due to medication noncompliance as opposed to treatment failure. It was

hypothesized that undetected poor compliance to Beta-blocker therapy may lead to unnecessary ICD implantations (Viskin & Halkin, 2009).

Although effective, there are associated risks of utilizing ICDs (Daubert et al., 2007). For instance, surgery can cause sympathetic activation, which can trigger arrhythmias, and anesthetic agents may prolong QTc intervals (Hobbs et al., 2007). Devices often need to be replaced several times through surgical procedures, increasing the risk for infections (Viskin, 1999). Patients may also experience painful and inappropriate shocks, multiple shocks, lead-related complications, and vascular blockages (Daubert et al.). Furthermore, ICDs are not a cure, as they do not prevent life-threatening arrhythmias (Schwartz et al., 1991). Thus, doctors often make recommendations in the hopes of preventing the onset of cardiac symptoms.

Since sudden cardiac death usually arises immediately before or after athletic activity, researchers have concluded that competitive sports likely increase the risk of cardiac arrest (Corrado et al., 2005). Although risk variations exist among LQTS genotypes, and individuals with LQT1 are at greatest risk for cardiac events during exercise, genotype-specific triggers are not mutually exclusive (Kapetanopolous et al., 2006). Therefore, exercise restriction is recommended, in varying degrees, in virtually all patients (Collins & Van Hare, 2006).

Prognosis. Previous researchers have also found that LQTS prognosis is based on a combination of genotype and phenotype. For example, the risk of a cardiac event is higher in patients with a history of aborted cardiac arrest, syncope, or those with a QTc interval prolongation over 500 milliseconds (Daubert et al., 2007; Priori et al., 2003). Priori et al. found that individuals with LQT2 had the highest mortality rate, followed by

LQT3 and LQT1. Interestingly, females with LQT2 had double the annual sudden death rate compared to males with LQT2, while males with LQT3 had approximately triple the annual death rate compared to females with LQT3 (Priori et al.). Another study concluded that probands, the first member of a family with a diagnosis of LQTS, typically have a more malignant course, although a selection bias may have contributed to increased event rates, since older probands were often recruited after a major cardiac event (Moss et al., 1991). Moss, Schwartz, Crampton, Locati, and Carleen (1985) also discovered that episodes of torsade de pointes or ventricular fibrillation are risk factors for sudden death. Viskin and Halkin (2009) also found that prognosis is based on compliance with Beta-blocker therapy and avoidance of medications that may prolong the QTc interval. Therefore, prognosis depends on clinical presentations and genetic variations, creating an inseparable link between genotype and phenotype.

Association with other disorders. LQTS has been correlated with sudden infant death syndrome (SIDS), the leading cause of death in infants (Arnestad et al., 2007). Arnestad et al. stated that life threatening arrhythmias caused by LQTS are one cause of SIDS. This study found that LQTS gene variants were associated with 9.5% of SIDS cases. Otagiri et al. (2008) found that nearly 10% of Japanese SIDS cases were associated with LQTS gene mutations. Additionally, since approximately 30% of LQTS genetic mutations have yet to be discovered, these findings likely underestimate the link between SIDS and LQTS (Arnestad et al.). The authors hypothesized that infants experience cardiac electrical instability due to bursts of sympathetic activity during sleep, a propensity for upper respiratory tract infections, and sleeping position (Arnestad et al.).

What researchers still need to study. Although LQTS researchers have made remarkable progress in the past few decades, there are still aspects of the syndrome that have yet to be discovered. For instance, although the genetics of the syndrome are better understood, more needs to be uncovered pertaining to disease expression and the relationship between genetics and clinical presentations (Collins & Van Hare, 2006). Once these factors are uncovered, gene therapies may be able to target specific mutations to correct the electrical abnormalities, prognosis will be better understood, and preventative measures will become clearer. Otagiri et al. (2008) stated that DNA tests may one day be able to screen infants for SIDS-associated mutations, preventing tragedy. Clinicians need to better understand risk stratifications and the associations between symptoms and prognosis to better treat patients and make appropriate recommendations (Kapetanopolous et al., 2006). It is still unclear what mechanisms are responsible for the age and sex risk differences in LQTS (Locati et al., 1998). Uncovering these relationships may shed light on treatments or preventative measures. Additionally, discussions pertaining to the pros and cons of widespread neonatal ECG screenings should continue (Arnestad et al., 2007). Although costly, standard screenings could save hundreds or thousands of lives, as most deaths due to LQTS can be prevented (Schwartz et al., 2000). For instance, Lawless and Best (2007) suggested that ECG screenings should be conducted on symptomatic athletes or those with an abnormal history or physical examination. Kapetanopolous et al. stated that all athletes with fainting episodes, a history of cardiac arrest, or a family history of sudden death at a young age are candidates for a diagnostic workup and should be evaluated. Furthermore, Corrado et al. (2005) stated that efforts should be made to increase the use of ECG screenings for all athletes,

even those who are healthy, in order to reduce the risk of untimely deaths. Hence, increased efforts to promote awareness and education about LQTS in communities, schools, and for doctors are needed.

LQTS Regimens

Restrictions. Due to the potential for life-threatening cardiac arrest, preventative measures are often taken, and restrictions are often placed on individuals with LQTS to reduce the likelihood of fatal arrhythmias. Since symptoms are often precipitated by physical exertion, such as water sports, and intense emotions, risk stratifications have been created for young athletes or individuals who are physically active (Kapetanopolous et al., 2006). Typically, all LQTS patients are restricted from competitive sports (Kapetanopolous et al.; Zareba & Cygankiewicz, 2008). This rule especially applies to LQTS patients who have suffered a cardiac arrest and/or a fainting episode (Kapetanopolous et al.). Specifically, activities involving a burst of exertion, adverse environmental conditions, progressive levels of exertion, excessive participation, or cardiac stress should be avoided (Maron, Chaitman, Ackerman, Bayes de Lina, Corrado et al., 2004). Schools can play a critical role in monitoring adherence to these recommendations and creating alternative athletic curriculums.

The acceptability of recreational sports participation, which involves less rigorous expectations and training, is graded on a relative scale, ranging from high to low intensity, based on levels of physical exertion (Kapetanopolous et al., 2006; Maron et al., 2004). For example, golf, bowling, or brisk walking are “probably permitted,” while more rigorous activities, such as basketball, soccer, or swimming, are “strongly discouraged” (Kapetanopolous et al.). In their study, Kapetanopolous et al. suggested that

limitation determinations be individualized after evaluating the risk of exercise, gene-specific triggers, personal preferences, and the emotional impact of the restrictions. Consequently, the risks of the activity should be weighed against the benefits of participation.

Additional restrictions, aside from athletics, have also been noted in the literature. For instance, individuals with LQT2 should avoid sudden loud noises, such as starting pistols, alarm clocks, telephones, or doorbells, as they are often triggers (Kapetanopolous et al., 2006; Schwartz et al., 1991). LQTS patients, especially those with LQT2, should try to avoid emotional stress, fright, or anger, as the activation of the sympathetic nervous system can lead to tachycardia (Kapetanopolous et al.; Schwartz et al.). Since distress and eustress are aspects of daily living, attempts to avoid or curtail emotional experiences may create heightened frustration and anxiety (Zareba & Cygankiewicz, 2008). Therefore, future studies should specify ways patients can successfully curtail emotional reactions. Medications that have been shown to prolong the QTc interval, such as certain antibiotics, stimulants, and antihistamines, should also be avoided (Kapetanopolous et al.). Lastly, foods or substances that increase sympathetic arousal, such as those containing caffeine or chocolate, should be avoided and can be monitored by designated school personnel (*Sudden Arrhythmia Death Syndromes Foundation*, 2008).

Additional needs. Since individuals with LQTS face the fear of cardiac symptoms, require daily treatments, and are restricted from activities that “healthy” people typically engage in, this population may benefit from additional services. The restrictions placed on individuals with LQTS, such as with athletic sports participation, may cause emotional stress (Kapetanopolous et al., 2006). Individuals with LQTS, especially youth, are asked

to give up activities that may impact their self-esteem and social enjoyment (Vetter, 2007). For those in whom the condition is diagnosed after training for athletics or being a part of competitive sports, it may be a cognitive shift in their concept of identity (Napolitano et al., 2006). Maron et al. (2004) stated that exclusions from school-structured physical education class may be associated with stigmatization and ostracism from peers. Moss et al. (1995) stated that family counseling may be required to direct the interests of the affected individual to less physically demanding and emotionally straining pursuits. Hence, schools should implement accommodations and crisis plans that address the social, emotional, and medical needs of the population, considering developmental appropriateness and student rights.

Zareba and Cygankiewicz (2008) suggested that patients and family members of those with LQTS receive education about the importance of medication and restriction compliance, along with frequently updated knowledge about drugs that can prolong the QTc interval. Viskin and Halkin (2009) recommended that patients and physicians be periodically reminded of the harms regarding unnecessary medication use. Lastly, patients with LQTS should receive cardiopulmonary resuscitation (CPR) training and own automated external defibrillators (AED) as an additional safety measure (Viskin & Halkin; Zareba & Cygankiewicz). These recommendations should be extended to schools and specific staff responsible for children with LQTS to maximize compliance and safety. Thus, a school-based assessment and checklist tool would aid in facilitating this process.

LQTS youth may also require school-based accommodations. For instance, Vetter (2007) indicated that families should work with schools and athletic coaches to find a

balance between restricted activities and those that allow patients to interact with their peers. Since personal and social stigmas may be associated with gym class prohibition, it is often useful for parents and school personnel to review physical education class requirements to create a plan to minimize ostracism (Maron et al., 2004). Equally important is the schools' recognition of the syndrome and the agreement that patients have the authority to cease athletic participation when necessary (Maron et al.). The Sudden Arrhythmia Death Syndromes Foundation (SADS) recommends that all LQTS patients be supervised during physical activity, a buddy system be implemented in which another individual is informed about symptoms and risks, and school personnel and students be instructed to call for help in an emergency and learn CPR (*Sudden Arrhythmia Death Syndromes Foundation*, 2008). It has also been recommended that family, friends, and neighbors be made aware of the condition, and every LQTS patient should wear an identification tag to notify emergency workers (*Long QT Syndrome*, 2008). Lastly, automated external defibrillators should be placed in schools for emergency cardiac treatment, and staff should receive training (*Long QT Syndrome*, 2008). A sample accommodation plan was developed based on guidelines from other chronic disorders, such as diabetes, and the steps to construct and implement comprehensive school-based LQTS accommodations are detailed in this manuscript.

Although these suggestions are potentially lifesaving, the literature does not indicate how to appropriately implement these accommodations while taking into account the likelihood for cognitive and emotional impact. For instance, making schools aware of a child's condition could create greater stigmas, in turn leading to emotional stress, which is often an LQTS trigger. Monitoring and individualization of curricula could lead to

ostracism (Maron et al., 2004). Adolescents may resent the assistance of classmates, resisting the buddy system. Youth may feel their confidentiality is violated or experience differential treatment due to others' fear or misunderstandings of the syndrome.

Therefore, although these recommendations are seemingly useful, they need to be implemented in ways that will maximize the likelihood of adherence and minimize the risks of social, academic, emotional, or psychological impact, such as following a school-based manual or procedural guideline, as provided in this manuscript.

Importance of Studying this Population

Unpredictability. Given the unpredictable nature of this condition, individuals may feel uncertain about their life expectancy and when a cardiac event may occur. Since patients are typically asymptomatic and feel healthy, it is difficult to predict when a cardiac arrest could happen. Additionally, since forewarnings for cardiac arrest, such as syncope or dizziness, do not always occur, patients and families may live in constant fear of the worst (Vetter, 2007). Although different risk stratifications have been associated with LQTS variants, they are not mutually exclusive, potentially making all triggers dangerous. Hence, although patients with LQTS are in control of their adherence to medical advice, they still ultimately live with the unpredictability of their fate. Medical adherence is therefore the primary way for the LQTS population to take control of their syndrome, making the syndrome more predictable and less fatal. Schools can offer support to adolescents with LQTS, providing counseling sessions focusing on feelings of uncontrollability and medical adherence.

Medical adherence. Medical adherence, especially with potentially life-threatening conditions, is critical (Garson et al., 1993). One study found that

noncompliance with previously effective LQTS treatment was a predictor of symptoms and sudden death (Garson et al.). Since the protective effects of Beta-blocker medications dissipate within a day or two of stopping medication, the danger for sudden cardiac arrest returns, reverting back to the highest risk category (*Sudden Arrhythmia Death Syndromes Foundation*, 2008). Permission to play athletic activities is often based upon the appropriate prescription of Beta-blocker therapy and compliance (Napolitano et al., 2006). Therefore, if medical regimens are not adhered to, but patients still engage in exercise activities, their risk of sudden death becomes elevated. Schwartz et al. (1991) stated that cardiac arrest is often precipitated by emotional or physical stress in otherwise healthy LQTS patients. Consequently, if these patients remain untreated, these triggers may eventually prove fatal (Schwartz et al.). School staff can monitor medication compliance and adherence to physical activity restrictions and recommendations, increasing the likelihood of safety and health.

Health beliefs. Given the risks associated with LQTS, it is critical to increase patients' health behaviors, which are actions taken that enhance or maintain good health (Taylor, Peplau, & Sears, 2000). Overall, it is critical for patients to take their daily medications, visit the doctor regularly for follow-ups, adhere to medical advice, seek additional consultations or opinions, and avoid foods and drugs that could induce LQTS. In general, LQTS patients should be encouraged to take an interest in their health, have an accurate understanding of the threats to their health, believe in their personal vulnerabilities, believe that they are able to reduce their medical threats through treatment adherence, and suppose that their actions will be efficacious (Taylor et al.). Maintaining these cognitions will increase adherence to medical regimens and ultimately lead to

happier, healthier, and longer lives. School staff, such as nurses, counselors, and psychologists can provide support and assess cognitions that lead to healthier beliefs and behaviors.

Adherence challenges. Adherence may be especially challenging for LQTS patients, as they often remain or become asymptomatic throughout the course of their treatment (Vincent et al., 2009). For instance, patients may believe that they are cured and begin questioning the need for continued medical treatment, resulting in skipping or stopping their medications (Vincent et al.). Often, health beliefs are related to one's perception of susceptibility to symptoms and the severity of the condition (Llewelyn & Kennedy, 2003). Additionally, patients typically weigh the benefits against the costs of taking health-related actions (King, 1984). If patients have positive cognitions about their vulnerabilities and the seriousness of the syndrome, believing that they are asymptomatic and therefore healthy and at minimal risk for cardiac arrest, poor compliance and reduced treatment adherence might result, greatly increasing risks. Alternately, since medication compliance is often related to the perceived necessity of the regimens, and LQTS cardiac symptoms do not always occur when medication is ceased, patients may not see the utility in the treatment (Kyngas, Kroll, & Duffy, 2000). Kyngas et al. stated that factors associated with compliance also include therapeutic motivation, daily experience of symptoms, and attitudes towards the illness. Furthermore, the extent to which patients attribute their symptoms as being in their control impacts their health behaviors (King, 1984). Since symptoms of LQTS are often unpredictable, it is possible that patients may give up on their medical regimens. Lastly, noncompliance associated with simple forgetfulness can be remedied with simple reminders, such as cell phone alerts (Viskin &

Halkin, 2009). Therefore, LQTS is a unique condition, as patients may feel otherwise healthy, do not have structural cardiac abnormalities, and may live symptom free.

Unfortunately, at the root of the syndrome is a dormant electrical instability which could prove fatal. Hence, it is critical for patients to receive education about LQTS and become periodically reminded of their risks and limitations in hopes of maintaining medical adherence. For adolescents with LQTS, schools can aid in education and guidance with medication adherence.

Differences Between Healthy and LQTS Populations

Although they may otherwise be symptom free, differences exist among LQTS and healthy populations. LQTS patients may think about the uncertainty of their future and lifespan. Living with a condition that could prove fatal may create feelings of fear, depression, or helplessness. Often patients are restricted from engaging in developmentally appropriate activities because physical exertion or activities could trigger sympathetic nervous system activation. Thus, children and adolescents with LQTS may become stigmatized or socially ostracized from their peers. School-based accommodations for youth, such as a buddy system, or the awareness of teachers, may make them feel different from others, potentially reducing their self-esteem. It is possible that others may treat LQTS patients differently, due to fears or misunderstandings of the syndrome. Furthermore, medical adherence is critical for this population, unlike healthy individuals whose life may not hang in the balance. Therefore, although this population may appear to be healthy, the social and physical restrictions, medical treatments, and accommodations made by schools, parents, family members, and friends may create

feelings of isolation and poor self-perceptions. Given the importance of schools in adolescent social and emotional development, accommodation plans can be implemented to maximize school connectedness and belonging, advocating for the needs of these youth.

Sparse Literature Exists

Although an abundance of LQTS medical literature exists, there are few studies pertaining to the psychosocial effects of living with LQTS. One recent study investigated the course of distress among adults undergoing LQTS genetic testing and found that disease-related anxiety scores were high among carriers (Hendriks, Hendriks, Birnie, Grosfeld, Wilde et al., 2008). The authors hypothesized that uncertain medical treatments, the need for lifestyle changes, threat of sudden death, and the genetic nature of the syndrome likely contributed to increases in anxiety (Hendriks et al., 2008). Another study assessed anxiety levels among a small sample of children with LQTS and found high scores on internalizing and externalizing behaviors, hypothesized to be a result of unresolved anxiety (Giuffre, Gupta, Crawford, Leung, 2008). These authors also found that parents of children with LQTS reported high levels of state anxiety (Giuffre et al.). The rates of depression in the LQTS population are currently not known, although researchers estimate that between 2% and 5% of the general adolescent population experiences at least one episode of major depression (Costello, Mustillo, Erkanli, Keeler, & Angold, 2003). Although unknown whether adolescents with LQTS are more prone to depression than the general population, it is suspected that they may experience psychological distress due to activity restrictions, medical regimens, and feelings of

uncertainty and difference. Future studies should investigate the rates of depression in adolescents with LQTS, ultimately providing prevention and intervention efforts, if needed. In a prospective follow-up study investigating parental adjustment to a child's diagnosis of LQTS, Hendriks, Grosfeld, van Tintelen, van Langen, Wilde et al. (2005) concluded that parents experience higher levels of distress and anxiety after a positive diagnosis and at 18-month follow-up compared to parents whose children were negative for LQTS. For instance, parents of children with LQTS reported sleeping with the doors open, being vigilant for symptoms, and having fears regarding medical noncompliance once their children entered puberty (Hendriks et al., 2005). In a qualitative study of coping among a sample of 8 Norwegian adults with LQTS, Andersen, Oyen, Bjorvatn, and Gjengedal (2008) found positive response to diagnosis, anxiety and worry, limitations and loneliness, and risk and existentiality as predominant themes. In this study, participants' anxiety and worry included uncertainty, unresolved emotions, and uncontrollable symptoms (Andersen et al.). Patients also spoke about worry after experiencing cardiac events, concern for other family members, limitations in daily life, fears regarding ICD functioning, others' lack of knowledge regarding the syndrome, fears regarding future pregnancy, and dissatisfaction regarding LQTS knowledge among healthcare providers (Andersen et al.). Given the results of these preliminary studies, it is perceived that individuals with LQTS are in need of intervention guidelines.

Unlike many other conditions, there is a lack of research to help families or patients cope with the condition. There is also a need for information pertaining to the self-esteem of children and adolescents with LQTS, given the potential for ostracism and social isolation. Since youth milestones, such as establishing friendships, engaging in

athletics, or school functioning may be harder to attain for these individuals, it is essential to uncover ways to make these periods progress smoothly. Specifically, given the tenuous development of adolescence, coupled with the social and physical restrictions that adolescents with LQTS face, this population could benefit tremendously from support from the psychological and school-based communities, such as the development of accommodation and crisis plans. Although intuitively, differences exist between the healthy and LQTS population, it is critical that we understand precisely how these disparities impact individuals with LQTS. It is through these explorations that the psychological and medical community will be able to implement better treatments and accommodations to assist these individuals in leading fulfilling lives.

Adolescents and Self-Esteem

Characteristics of Self-Esteem

Self-esteem definitions. Throughout the literature, self-esteem (SE) has been defined in many different ways by many different researchers. Some theorists claim that one's self-image, or the subjective perception of oneself, is based on measurable quantities, such as genetic attributes, acquired material possessions, and quantifiable products of work (Bailey, 2003). The individual places value upon these assessments, influenced by one's personal schema, or a collection of beliefs about oneself, others in the social environment, or by greater society (Bailey). These subjective views are also based on sense of self over time, relative in comparison to others (Sukumaran, Vickers, Yates, & Garralda, 2002). Hence, although people may look objectively at their qualities

and possessions, their determination of self-worth is ultimately based on subjective values and ideals.

Other theorists believe that SE consists of mental pictures of one's attributes, including physical appearance, feelings, desires, and experiences (Bailey, 2003). Therefore, SE may rest in the eye of the beholder, rather than in objective criteria. Taking this subjective stance, Searcy (2007) stated that SE consists of an individual's feeling about himself or herself, implying that one's emotions may impact perceptions of self-worth and adequacy. Corroborating with this belief, another theorist indicated that SE could be measured by simply answering "how much do I like myself?" (Hendel, 2006). Thus, this question is based on personal opinion rather than quantifiable data.

SE has also been conceptualized as an overall evaluation of one's competency, based on self-efficacy, worth, and feelings of personal value (Cast & Burke, 2002). For example, Santrock (2001) stated that SE is the global evaluative dimension of the self. Again, one's personal evaluations may not be grounded in objective criteria, but are relative to personal norms, values, and thoughts (DuBois, Bull, Sherman, & Robert, 1998). This view of SE also highlights the overall summation of one's self-evaluations, rather than an analysis of component parts. Alternately, other researchers believe that SE is better captured through a breakdown of personal qualities.

Types of self-esteem. Consistent with the existence of varying SE definitions, studies have also identified multiple types and variations of SE. Throughout the literature, researchers have debated whether SE is better captured as a global "gestalt" construct or as multidimensional components. For instance, Wilkinson (2004) stated that a view exists in which SE is the evaluative component of the self-concept and is more global than

independent characteristics. From this view, SE is therefore better measured by assessing one's general feelings of self-satisfaction. Alternately, others evaluate SE by looking at the characteristics that underlie the construct (Wilkinson). For instance, DuBois et al. (1999) stated that an exclusive focus on global self-worth yields limited understanding of one's SE, as it does not tap into sensitive developmental and contextual influences, limiting the ability to identify SE profiles and characteristics. Furthermore, multidimensional aspects of SE may be independent of global SE, adding critical pieces of information when assessing perceptions of self-worth (DuBois et al., 1998).

For those who measure SE by analyzing its component parts, there is unfortunately little uniformity regarding what those modules should consist. For instance, Searcy (2007) stated that SE can be categorized into explicit SE, which is expressed feelings of oneself, implicit SE, which are automatic thoughts of worth, optimal SE, which are non-contingent positive feelings of self-satisfaction, and global SE, which are emotions and feelings of likeability. Quatman and Watson (2001) included eight SE categories when measuring gender differences in adolescent SE: personal security, home/parents, peer popularity, academic competence, attractiveness, personal mastery, psychological permeability, and athletic competence. In another study, a cluster analysis indicated that peers, school, family, appearance, and sports were associated with youth SE (DuBois, Felner, Brand, & George, 1999). Shapka and Keating (2005) found that appearance, scholastic competence, job competence, close friendships, and social acceptance were significantly associated with overall self-worth in adolescents.

A widely used measure of SE, the Perceived Competence Scale for Children (Harter, 1982), includes four distinct SE domains: cognitive competence, scholastic

competence, physical competence, and global self-worth. Harter (1982) included these domains, stating that global measures do not adequately capture all SE elements (Harter, 1990). In the Harter questionnaire, global SE is a separate measure, distinct from the other subscales, as overall SE was found to be distinct from other areas of competence (Sukumaran et al., 2002). Harter also stated that general SE measures do not tap into concerns fundamental to the developmental period of children and adolescents (Harter, 1982). For instance, increases in academically demanding subjects, the emerging importance of acceptance in peer groups, the value of athletic ability, and the ability to look introspectively are SE domains that are critical to youth SE (Harter, 1982). In later iterations of the Harter measure, domains including physical appearance and behavioral conduct were added to the instrument to increase its validity (Harter, 1985).

Given the varying conceptualizations of SE, the constructs lack consistency in their definitions, frequently making them appear ill defined and nebulous. Often, researchers do not give the rationale as to why specific SE components have been included, making the variables appear arbitrary. Furthermore, SE module definitions often overlap, making distinctions unclear and redundant. In order to make comparisons between findings, future research should attempt to systematically define the constructs and create nuanced terms to identify the varying SE dimensions.

Development and maintenance of self-esteem. Another critical component of SE is the way in which it is developed and maintained. Researchers have indicated that there are multiple determinants of SE, including associations with others, engaging in activities, and hearing others' opinions (Searcy, 2007). Specifically, social groups, peer groups, family, and friends have been found to impact SE (Searcy). Often, feelings of

achievement arise when one receives positive reinforcement and social rewards from others through participating in activities and mastering skills (Searcy). A cyclical process emerges where praise from accomplishments is used as evidence for future competence, which further feeds into feelings of self-worth (Gecas & Schwalbe, 1983). SE is thus a dynamic construct, changing based on environmental expectations, successes, and expectations (Baldwin & Hoffmann, 2002).

SE is also maintained when people seek opportunities to verify their self-conceptions and avoid circumstances that contradict their beliefs (Cast & Burke, 2002). Hence, a cyclical process emerges where SE becomes maintained by filtering out stimuli that run counter to one's beliefs and by increasing time in reinforcing environments and social situations (Cast & Burke). Although sometimes unaware of this process, people simply engage in activities and spend time with people that make them feel good about themselves. Harter and Monsour (1992) also found that positive self-characteristics are frequently considered the core constructs of one's personality, while less desirable characteristics are considered peripheral. Consequently, focusing on the positive and minimizing the negative maintains SE. For instance, since adolescents with LQTS are often restricted from competitive sports, they may avoid friendships with athletes due to feelings of inferiority, preferring individuals who are able to reinforce other abilities, such as artistic skills.

Predictors and influences of self-esteem. Just as a number of factors aid in the development and maintenance of SE, there are also factors that influence SE both positively and negatively. These are situational and dispositional factors that impact self-evaluations, including contextual and developmental influences that are often interrelated

(DuBois et al., 1999). For instance, according to identity theory, SE becomes verified when social situations confirm one's personal perceptions (Cast & Burke, 2002).

Therefore, circumstances that corroborate beliefs of personal worth may lead to increases in SE, while disconfirming states may reduce ratings of SE. These results indicate that SE may be influenced by situational variables, not internal dispositional characteristics.

In their research, Quatman and Watson (2001) found that global SE is strongly influenced by parents and home life, personal security, academic performance, and being sure of oneself. Substantiating the association among families and SE, Hendel (2006) found that boys of alcoholic parents tend to have lower ratings of SE, possibly due to reduced parental interactions. Laursen, Furman, and Mooney (2006) found that supportive mother-daughter relationships were related to adolescents' global self-worth, and social support in friendships was associated with social acceptance, friendship competence, and romantic competence. Likewise, Macek and Jezek (2007) found that perceived relationships with parents and peers are predictors of SE. Specifically, researchers found a strong correlation between SE and warmth and trust of the parent of the same sex (Macek & Jezek). Finally, Rubin et al. (2004) found that greater perceived parental support and friendship quality predicted higher ratings of global self-worth and social competence, fewer internalizing and externalizing disorders, and less peer victimization. Hence, along with peer friendships, supportive and trusting parent-child relationships in LQTS patients may aid in the development and maintenance of positive self-worth. Particularly in youth who are restricted from peer activities, familial relationships may bolster feelings of confidence and competence that are not achieved through social interactions.

As a dispositional characteristic, depression may also be a contributor to low SE, as it often involves social and interpersonal dysfunction, which may impact one's personal image (Hendel, 2006). Another study investigating the association between SE and adolescent psychiatric patients found that older adolescent boys with emotional and psychosomatic disorders, including depression and anxiety, were especially prone to reductions in scholastic, athletic, social, and global SE (Sukumaran et al., 2002). In another study, Hall-Lande et al. (2007) found that social isolation is associated with depression, suicide attempts, and low SE. In this research, protective factors included family connectedness, academic achievement, and school connectedness (Hall-Lande et al.). For females, family connectedness was most helpful in mediating the negative effects of social isolation, whereas for males, academic achievement and school connectedness were most protective (Hall-Lande et al.). Although research is needed pertaining to the association between LQTS and depression, adolescents with LQTS may be prone to internalizing disorders, including depression, due to activity and social restrictions. The life-threatening nature of the condition, coupled with its chronic course, may also increase feelings of hopelessness and despair, which are related to reduced SE.

Age may be another predictor of SE. For example, boys' SE often declines between the ages of 14 and 16, potentially due to changes in physical maturation, emotional development, and schooling transitions (Hendel, 2006). For females, SE increases until age 12, drops until 17, and then rises again (Baldwin & Hoffmann, 2002). Hence, SE has been found to be a dynamic rather than static construct, supporting a developmental perspective (Baldwin & Hoffmann). Overall, many of these categories are interrelated, as negative experiences in one domain, such as depression, may adversely

impact another domain, such as peer relations, which further increases initial negative feelings (DuBois et al., 1999; Hall-Lande et al., 2007). These cyclical relationships therefore serve as another SE influence, feeding into positive and negative self-perceptions. For example, the restrictions placed on adolescents with LQTS may increase patients' feelings of hopelessness and depression, thereby leading to further withdrawal, perpetuating poor social functioning. For this reason, breaking these self-perpetuating cycles can allow individuals with LQTS to alter their behaviors and negative thought patterns, potentially improving their SE.

A substantial portion of the SE literature has investigated the relationship between SE and gender. In their explorative study, Quatman and Watson (2001) found that boys scored higher than girls on six of eight SE domains, including home/parents, personal security, academic competence, attractiveness, and personal mastery, and on global SE. Harper and Marshall (1991) found that girls had significantly lower SE scores than boys, including more problems with interpersonal relationships, personal adjustment, physical development, health, and family issues. Thomas and Daubman (2001) found that females reported lower SE, predicted by cross-gender friendship quality, interpersonal reward, and relationship strength. Finally, Chubb, Fertman, and Ross (1997) concluded that adolescent females reported significantly lower SE than males consistently throughout high school, implying that these divergences occurred even before ninth grade.

Male and female global SE ratings may also be qualitatively different (Knox, Funk, Elliot, & Greene Bush, 1998). In their study, Knox et al. found that although male and female global SE scores did not differ significantly, females' SE ratings correlated with their self-report of satisfaction and happiness, prospects of accumulating wealth and

establishing financial independence, having high educational and occupational hopes, anticipated establishment of relationships, and physical appearance. Alternatively, males' global SE was only correlated with self-confidence in social situations (Knox et al.). Thomas and Daubman (2001) stated that females are socialized to value relationships more than boys, causing friendship quality to affect the SE of girls more, whereas boys report a lower need for intimacy. Results also indicate that low SE in males tends to correlate with athleticism and academics, while for females it translates to academic and social functioning (Knox et al.). To corroborate this statement, Shapka and Keating (2005) found that boys had higher perceptions of physical and athletic competence, whereas girls had higher perceptions of social and friendship competence. Hence, even when looking at global SE, it may be telling to assess the different factors that load into the score. For adolescents with LQTS, it is therefore important to assess multiple SE domains, including social and school functioning, in order to obtain a valid assessment of the overall self-perceptions of both genders. Although it is possible that gender differences exist in the current population of focus, it is premature to hypothesize what the differences may be.

The discrepancies among male and female SE may be due, in part, to the norms that society and families dictate for each gender (Harper & Marshall, 1991; Knox et al., 1998). For instance, it is more socially sanctioned for females to acknowledge having physiological symptoms, such as headaches or stomachaches, reducing ratings of social security (Quatman & Watson, 2001). Likewise, following stereotypical male expectations, boys often indicate possessing leadership roles and taking risks, raising personal mastery scores (Quatman & Watson). Hendel (2006) stated that most self-

esteem issues in males occur when they do not fit the stereotypical prototype, such as being athletic, strong, and independent. The media also promote images of physical beauty and physical attractiveness for females and wealth and aggressiveness for males, creating nearly unattainable standards and ideals from which to make value judgments (Harper & Marshall). For example, mid-adolescent females tend to value physical appearance, in which perceiving themselves as unattractive correlates with lower SE ratings (Knox et al.). In line with society's expectations, one study found that boys rated their educational-vocational future as being associated with their SE, while girls' SE was correlated with problematic relationships, courtship, health, and physical development (Harper & Marshall). Furthermore, society's encouragement of females to resolve discrepant roles and attributes may place stress on women (Harter & Monsour, 1992). Therefore, reductions in SE may ensue when social norms are violated. Gender-related SE is therefore a function of societal pressures, familial expectations, and personal beliefs.

Consequences of self-esteem. Ratings of high and low SE also relate to proximal and distal outcomes. For instance, Harter (1990) indicated that high ratings of SE correlate with higher quality of life ratings. Alternatively, low ratings of SE are correlated with emotional and behavioral disorders, including anxiety, suicidal behavior, delinquency, eating disorders, and depression (Harter). Quatman and Watson (2001) indicated that SE is related to quality of life, motivation, depression, and resiliency. In their study, DuBois et al. (1999) also concluded that self-derogatory adolescent profiles were predictive of increased emotional and adjustment problems. In a similar study, researchers found that higher reported levels of global SE were correlated with higher

scores on adjustment measures, fewer internalizing and externalizing disorders, and higher ratings of academic achievement (DuBois et al., 1998). Ybrandt (2007) found that positive self-concepts were protectors against behavior problems, while negative self-ratings were associated with internalizing disorders, which later predicted aggressive and delinquent behaviors. Hendel (2006) stated that individuals with low SE often exhibit a need to win, perfectionism, self-criticalness, and a tendency to blame others for their downfalls. In particular, males with low SE are prone to befriend peers with negative behaviors, succumbing to negative peer pressure, acting aggressively, and breaking rules. Fitting in with any crowd, even the “bad” crowd, is a way to increase the level of belonging (Hendel). Children with low SE also have a tendency to develop a sense of hopelessness, which in turn may lead to further reductions in SE (Hendel). Therefore, if the cycle is not broken, the effects of low SE may lead to internalizing and externalizing disorders, including depression, self-harm, delinquency, aggression, anxiety, somatic complaints, and withdrawal (Hendel; Ybrandt). Furthermore, Ybrandt stated that internalizing and externalizing problems tend to co occur and influence each other, creating even greater risks. In order to establish causal relationships among SE and psychosocial functioning in adolescents with LQTS, future research should employ prospective longitudinal studies that investigate SE from the time of diagnosis forward.

Overall, the literature is unclear whether those who have lower SE ratings are prone to psychological disorders, or if reductions in SE then increase vulnerabilities for mental health conditions. Given these correlations between SE and positive and negative outcomes, it is critical for researchers to target individuals with low SE to implement prevention or treatment programs. For instance, if individuals with LQTS tend to fall

prey to negative peer comments surrounding their syndrome, it would be useful to aid them in discovering positive friendships and maintaining positive self-cognitions. Assisting in the development and maintenance of positive SE would then serve to promote adaptive behaviors which would increase SE. As a preventative measure, individuals could be screened, and those LQTS patients whose results are associated with negative outcomes could be targeted for intervention (DuBois et al., 1999). On the other hand, researchers should continue to investigate protective SE factors, such as family connectedness, involvement in after-school activities, and service learning, to promote self-enhancing behaviors (Hall-Lande, 2007). The ultimate treatment or prevention program would aid in the creation of a self-propelling cycle of positive self-perceptions. With this information, there is a potential to increase protective factors rather than simply decreasing risk factors. The comprehensive school-based assessment and accommodation guidelines provided in this manuscript will assist schools in identifying adolescents who may be at-risk for low SE, and support their healthy psychosocial functioning.

Adolescent Self-Esteem

Adolescent development. Volumes of literature point to developmental shifts that transpire in adolescence, including biological, cognitive, and social metamorphosis (Santrock, 2001; Steiner, 1996). Adolescence generally consists of three phases, including early adolescence (10 to 13 years), mid-adolescence (14 to 17 years) and late adolescence (18 to 20 years) (Steiner). During these periods, developmental changes transpire, including marked changes in physical appearance, such as sex characteristics and accelerated growth brought upon by puberty, cognitive changes, including the ability

to think in abstractions, and identity shifts, such as increasing one's roles and independence (Steiner). Furthermore, adolescents typically expand their peer relationships to include opposite-sex partners and sexual relationships (Steiner). Lastly, adolescence is often characterized by school transitions, including moving to middle and high school, increasing academic pressures, and shifts in family dynamics, such as wanting more privacy and autonomy from their parents (Steiner).

Adolescence is a time of transition from dependent childhood to independent adulthood, characterized by a desire for autonomy and an increased value placed upon peer social support (Hall-Lande et al., 2007; Harper & Marshall, 1991). Peers are children or adolescents who are about the same age or maturity level (Santrock, 2001). More than any other age group, adolescents are typically concerned with their self-image and what others think of them, and the peer group takes center stage in developmental importance and parental influences slowly decrease (Hall-Lande et al.; Harper & Marshall). During adolescence, one's self-concept and identity formation are largely based on peers' perceptions of interpersonal skills, physical attributes, and achievement (Macek & Jezek, 2007). Hence, in order to attain higher social status, peer conformity and peer pressure often result (Macek & Jezek).

Shifts in adolescent self-esteem. Along with these developmental metamorphoses, the variables that impact SE also shift, increasing and decreasing in value. For instance, one of the hallmarks of adolescent maturation is an increased differentiation in one's self-concept and role-related selves (Knox et al., 1998; Quatman & Watson, 2001). During adolescence, one's self-descriptors, role attributes, and trait labels increase, creating more domains from which to base self-worth and SE (Harter & Monsour, 1992). To elucidate

this point, Harter and Monsour found that 7th, 9th, and 11th graders' self-descriptors included differentiated aspects of their identities, including self in classroom, with friends, with parents, and in romantic relationships. For example, an adolescent could create multiple selves, describing themselves as a loyal friend, a flirtatious partner, and a conscientious student (Harter & Monsour).

This increased multidimensionality in adolescent SE may be a result of the burgeoning cognitive ability to think abstractly, including how one is viewed by others (Harter, 1990). These cognitive abstractions allow the adolescent to integrate multiple attributions into a unified and coherent theory of his or her personality (Harter & Monsour, 1992). On the other hand, maturation is a slow process, and adolescents' abstract and hypothetical views of the self may be imprecise and implausible, leading to discrepancies among one's "true" self and hypothetical self (Knox et al., 1998). To corroborate this theory, one study found that young adolescents are only able to view themselves as one abstraction, middle adolescents are able to compare their varying attributes, but are not able to resolve internal conflicts when contradictory self-images arise, and it is not until late adolescence that individuals are able coordinate, resolve, and normalize discrepant self-views (Harter & Monsour). Therefore, for the majority of adolescence, individuals are unable to undergo the cognitive shifts necessary to resolve discrepant self-concepts.

Socialization pressures to integrate diverse roles and attributes also increase in adolescence, creating added demands and expectations to become a unified "self" (Harter & Monsour, 1992). Identity confusion may emerge when conflicts among self-images exist, including whom one is versus whom they would like to be (Harter & Monsour).

For instance, adolescents with LQTS may aspire to play competitive sports and become the star athlete, although the reality of their condition would prohibit the realization of that dream. Identity confusion may therefore emerge due to social and athletic restrictions, which are critical to adolescent development. Given the demands of society for differentiation and the cognitive inability to resolve self-discrepancies, there is the potential for adolescents with LQTS to experience role contradictions and dissonance, potentially leading to SE reductions (Harter & Monsour).

Functions of adolescent self-esteem. Adolescent SE is associated with multiple tasks, such as acquiring a better understanding of oneself, making sense of developmental changes, comparing oneself to peers, and aiding in social acceptance. Hence, peer groups often function to provide adolescents with information as to whether they are better than, as good as, or worse than their peers (Santrock, 2001). Adolescents' SE is also often based upon acceptance in peer groups (Burk & Caste, 2002). Verifying one's identity in a social group reinforces role-related behaviors and increases SE, making the adolescent feel accepted and liked (Burk & Caste). For instance, DuBois et al. (1998) stated that peer-oriented SE can be maintained through friendship formation activities which display common interests, such as extracurricular activities. Additionally, adolescents, especially females, frequently develop self-views through the perceived appraisals of others, demonstrating their competence by visibly displaying material possessions, increasing their external evaluations (Knox et al., 1998). In order to achieve others' positive perceptions, individuals often seek relationships that enhance their views of worth and efficacy and avoid interactions that do not (Burk & Caste). For adolescents with LQTS, restrictions from co curricular and athletic activities, receiving school-based

accommodations, and experiencing frequent medical events may create feelings of difference, hindering the task of social acceptance, potentially reducing SE. This population may therefore be unable to avoid those situations that diminish their sense of self-worth, perpetuating negative self-perceptions. Alternately, schools can maximize the likelihood of healthy SE by minimizing feelings of difference, such as allowing the child to take medications during breaks between class, providing them with noncompetitive gym class alternatives, and helping the child with medication adherence, reducing the chances of a cardiac event.

Adolescent peer relationships have been found to provide protective benefits, such as a sense of belonging, psychological support, emotional connectedness, and an outlet for trust (Hall-Lande et al., 2007). Peer acceptance is also an avenue to achieve identity formation and individuation (Sukumaran et al., 2002). For example, adolescents who report having close peer relationships tend to have higher ratings of emotional well-being and resilience and decreased levels of suicide risk (Hall-Lande et al.). In another study, supportive interpersonal interactions among cross-gendered relationships were predictive of higher SE, as adolescence is a time of relationship expansion and exploration (Thomas & Daubman, 2001). Research has also found that adolescents who associate with the popular crowd tend to have higher ratings of SE, indicating that group status is also associated with increased feelings of self-worth (Searcy, 2007). Furthermore, Fenzel (2000) concluded that peer support and social competence diminish strain during social and academic transitions, reducing stress in times of change while increasing self-worth. Hence, adolescents who have close relationships when transitioning to a new school seem to adapt more readily. In addition to SE, popularity

and peer acceptance often lead to job attainment, leadership roles, and recognition, further increasing SE ratings (Quatman & Watson, 2001). Adolescent friendships also serve multiple functions, including intimacy, companionship, nurturance, stimulation, physical support, social comparison, encouragement, and leadership (Jarvinen & Nicholls, 1996; Sullivan, 2001). Being sincere, having status, being responsible, pretending to care, entertaining others, and being tough were six behaviors believed to lead to successful peer relationships (Jarvinen & Nicholls). Adolescents with LQTS may have difficulty attaining peer acceptance due to real or perceived differences from healthy peers. For instance, limitations in co curricular activities may reduce opportunities to establish friendships, and school-based accommodations may highlight the need for medical attention. Furthermore, school-based activity limitations may leave LQTS patients with less in common with popular peers, reducing their chances for esteemed social status. As there are numerous benefits associated with social camaraderie, limited peer interactions may lead to feelings of loneliness and inadequacy. School staff can therefore help support the SE of adolescents with LQTS by providing accommodations to maximize school involvement and minimize feelings of isolation.

To become a member of peer and social groups, adolescents often abide by the gendered norms of society, since gender role socialization is a criterion by which to be judged (Harper & Marshall, 1991). For this reason, adolescents often conform to others, adopting the attitudes or behaviors of others due to real or imagined pressure to fit in (Santrock, 2001). Often, concerns about not being a part of a social group will increase conformity, whether for positive or negative behaviors (Santrock). For adolescents with LQTS, conformity, such as engaging in competitive athletics and fully participating in

school-based activities, may not be possible to attain. Furthermore, leaving class to take medications or receiving school-based accommodations may cause students to stand out, rather than blend in with other classmates. Hence, these differences may create feelings of being an outsider in one's own school, reducing SE ratings. Multiple theories point to reasons why peer group formations and social acceptance are so important in the development of adolescent SE, including symbolic interactions, social comparisons, and cognitive attributions.

Self-Esteem Theories

Symbolic Interactionism

Theoretical views. One theory that aims to explain the development and maintenance of SE is the symbolic interactionist view. Coined by Herbert Blumer in 1937, symbolic interactionism (SI) is a sociological theory in which one's "truth" is relative, since reality is constructed via interactions with the environment (O'Brien & Kollock, 1997; Schellenberg, 1990). Truth is produced through ongoing interactions and personal interpretations of social situations, a product of the context in which it was acquired (O'Brien & Kollock). Therefore, individuals classify and interpret data through self-constructed lenses which are culturally and historically bound, rather than through systematic quantitative methodologies (O'Brien & Kollock). Our perceptions also vary across the age span, as people's roles, interactions, and values change throughout development (Russell, 1984).

This theory states that human behavior is shaped by the meanings ascribed to situations and symbols, which emerge through interactions among individuals and

contexts (Russell, 1984). Our opinions of reality determine subsequent courses of action as well as what images and symbols we mentally store and retrieve (O'Brien & Kollock, 1997). Therefore, incidents do not inherently promote or reduce feelings of self-worth, but individuals assign symbolic meanings and abstract reasoning based on personal values. These perspectives then determine how people will act and feel as a consequence. As an example, for LQTS patients, restrictions from school-based activities are not inherently negative, but subjective values of importance ascribed to the activity and to peer conformity determine the impact on SE.

SI indicates that humans' behaviors are motivated by the attainment of rewards and avoidance of punishment, which are subjectively valued (O'Brien & Kollock, 1997). Furthermore, symbolic activity, such as words, language, and gestures, mediates the stimulus-response interaction, since it allows us to assign meanings, remember, plan, think, verbally communicate, transmit culture, and have vicarious experiences (O'Brien & Kollock). Therefore, although the focus is on observable behavior, the causes of actions are non observable interpretations (O'Brien & Kollock). Thus, self-esteem is impacted by the interpretations of abstract symbols, such as the words of others, where positive meanings increase self-worth and negative interpretations lead to SE reductions. SI would also contend that individuals seek environments that provide positive environmental feedback and avoid ones that do not, enhancing SE.

SI also points to the notion that people develop their individuality and sense of self through socialization, creating social contexts to shape collective standards and organize reality (O'Brien & Kollock, 1997). This is accomplished when people perceive the meanings of their behavior by surmising how it is viewed by others (Russell, 1984).

Even the term *individualism* is a socially constructed value, as it determines appropriate behavior within society (O'Brien & Kollock). SI therefore espouses the theory that social reality is created through the value-laden interpretations of the actions of social beings. Consequently, external social cues that adolescents are often conscious of, such as physiological characteristics and clothing, aid in the determination of one's acceptability in society through the values assigned to these culture-laden symbols. Therefore, social cues that differentiate healthy adolescents from those with LQTS, such as school-based accommodations and social restrictions, may lead patients to feel less accepted.

James Horton Cooley. One of the most notable SI theorists, James Cooley (1902), believed that self-concepts are based on the imagination of the reflections of one's appearance to others, otherwise known as the looking-glass self (Cooley, 1983; Lundgren, 2004). Individuals perceive in others' minds their face, body, dress, actions, character, manners, and aims, and are affected by subjective interpretations, otherwise known as reflected appraisals (Cooley; Gecas & Schwalbe, 1983). Hence, Cooley believed that the self represents one's perceptions of how one is viewed by others, a reflection of others' observations and evaluations, a metaphor for a mirror's reflection (Harter, 1995). This theory would claim that individuals with LQTS surmise what peers think of their syndrome and alter their self-representations based on their deductions.

Cooley's model purports that three elements exist when glancing in social mirrors: imagining one's appearance to others, imagining the judgments of others, and experiencing feelings based on thoughts of others' judgments (Cooley, 1983). Individuals' reactions are not mechanical reflections, but are impacted by the importance subjectively ascribed to the reflected impressions (Cooley). Therefore, after imaging

oneself in another's mind and ascribing value to it, it is common to share the judgments of the other person, changing and shaping SE (Cooley). Self-feelings, such as self-worth and SE, are therefore based on one's attitude and thoughts towards the attributions of the other person, which become an external source of self-worth (Gecas & Schwalbe, 1983). If individuals perceive others' views as positive, self-evaluations increase (Lundgren, 2004). Alternately, if individuals interpret others' views as looking down upon them, negative feelings ensue, whether from actual appraisals or merely perceptions (Lundgren). This view assumes that self-feelings are largely dependent on society as people spend a good amount of time in the minds of others (Scheff, 2005). This theory also points to the importance of the values' of others' opinions. In other words, Cooley's theory stated that SE is more likely to be affected when the people who are making the judgments are well-regarded. For instance, one study found that people's inclinations to accept or reject feedback vary depending on relationship closeness, emotional reactions, and feedback valence (Lundgren). Hence, peers would likely influence the SE of adolescents with LQTS due to the importance ascribed to them during this developmental period, especially if the peers are deemed popular.

Cooley and adolescent self-esteem. Children, especially, are able to make connections among their actions and the perceptions of their influence on the actions of others (Cooley, 1983). Behaviors that are perceived as making a positive impact are continued, while those with no effect or negative outcomes are discontinued (Cooley). Differentiated selves also begin to arise when these young "performers" perceive that different behaviors are acceptable in different settings (Cooley). As children age, they

also begin to desire that the internal states of others change, signifying acceptance and approval (Cooley).

Since the literature points to the impact of peer relationships on SE, it is likely that adolescents will either alter their self-appraisals to reflect the perceived judgments of others or change their behaviors to achieve positive self-images in the eyes of the valued evaluators (Gecas & Schwalbe, 1983). It is therefore reference groups, such as peers, that become the “mirrors” that reflect images of the self, establishing SE and self-awareness (Gecas & Schwalbe). Since individuals with LQTS are not often with others who have the condition, their comparators are healthy individuals, which may negatively impact their SE. Additionally, since those with LQTS are often restricted from changing their behaviors to achieve positive self-images, such as engaging in sports, changing their self-appraisals to match the judgments of others may be detrimental to their SE. Therefore, schools can play an important role in modifying the curriculum to allow students with LQTS to engage in behaviors that make a positive impact on their SE.

The concept of the looking-glass self also aids in the explanation of adolescent social conformity. Since, as research has indicated, peer acceptance is critical for normal adolescent development, it is likely that youth will please their “audience” by “performing” in ways that will achieve positive self-reflections. Since reputation, accountability, and relationships are based on the perceptions of others, it is critical for adolescents to constantly monitor their public behaviors, always acting fittingly (Tice, 1992). Youth with LQTS may therefore exert themselves to conform, such as participating in athletics when they should not, adding to the challenge of adhering to

medical advice. Moreover, engaging in activities that increase symptoms can lead to feeling different from others, further impacting SE.

In addition to perceptions of self-worth, Cooley held that a continuum of pride and shame arises from imagined judgments and self-monitoring (Cooley, 1983). Furthermore, he believed that global SE is comprised of the ratio between one's successes to one's aspirations towards success (Cooley). Thus, people place a value on the importance of achieving their accomplishments and weigh that against what they have already completed. Often, these aspirations are compared to the accomplishments of others, in our social realities, indicating the impact of social comparisons and SE (Goethals, 1986). For individuals with LQTS, the accomplishment of developmentally appropriate goals, such as successfully competing in sports and conforming to peers, may not be achieved. Therefore, this population may be at risk for reduced global SE, as their success and aspirations may be discrepant. School systems could support adolescents if they were well informed about the condition and sensitive to the relevant issues, such as collaborating with medical professionals and devising recreational athletic activities for the student to engage in. Advocating for the needs of adolescents with LQTS can also help increase their school connectedness and create feelings of empowerment.

George Herbert Mead. George Herbert Mead believed that reality is socially constructed through societal interactions, and symbolic communication enables individuals to assess their self-worth (Lundgren, 2004; Mead, 1962; Russell, 1984). Additionally, Mead (1962) held that there are distinctions among multiple parts of the self, including one's objective tangible body and the experience of one's self. The body can be conceptualized as the "self object," a physiological organism without memory or

imagination (Mead). On the other hand, the self is defined as one's indirect experiences through the standpoints of members of the same social group (Mead). Likewise, the "I" is one's conscious self-perceptions and reactions to others' views while the "Me" is a reflexive social object (Mead).

For Mead, the self is a social structure, created through public experiences, actions, consequences, and the product of efforts (Gecas & Schwalbe, 1983; Mead, 1962). Furthermore, the self is able to internalize the attitudes of others within a social environment and communicate accordingly, perpetuating the social process through symbolic language (Mead). As a critical component of Mead's theory, the self is able to engage in perspective-taking and role-taking by consciously observing oneself (Lundgren, 2004; Russell, 1984). These observations are made from the standpoint of specific people with whom one is interacting and a generalized imagined audience, allowing for self-assessments in comparison to social norms (Lundgren; Russell). Hence, Mead would likely state that for patients with LQTS, SE is derived from internalized perceptions of others and conscious feelings of self-efficacy.

Mead and adolescent self-esteem. In corroboration with the SE literature, Mead indicated that individuals have multiple parts to themselves, which get expressed differently in different relationships (Mead, 1962). Just as there are multiple aspects of one's self-worth, there are also different selves, depending upon what the social setting calls for, including public and private identities (Mead). This can be related to the differentiated social roles of adolescents, such as being a talkative friend, a studious student, and a loyal daughter. For example, personal secrets shared with a friend may not be expressed to a parent. As Harter (1995) indicated, children's' and adolescents' self-

competence is not a global construct, but a modular entity that can have different ratings for each component. Therefore, while individuals with LQTS may have reductions in certain aspects of their SE, such as social and school functioning, other areas of their self-perceptions may be intact, such as physical or behavioral conduct ratings.

Mead (1962) also stated that people experience self-consciousness when they perceive others' attitudes as attempting to change their behavior. In other words, individuals begin to question their behavior when they perceive that others are disapproving or critical. As the adolescent SE literature has indicated, this type of discrepancy between one's behavior and the opinions of others' often reduces SE (Knox et al., 1998). One of the risks that adolescents face when imagining the thoughts of others is that their comparisons can be skewed and faulty, leading to unnecessary SE reductions. Therefore, youth should be encouraged to systematically evaluate objective criteria when assessing their self-worth, improving their reality testing and reducing faulty assumptions. For instance, youth with LQTS may attribute their feelings of loneliness to their condition, discounting other characteristics, such as their personality, sociability, and friendliness towards others. Furthermore, adolescents with LQTS may imagine that their peers disapprove of their condition more than they actually do, leading to critical evaluations of peer interactions and perceptions of others' thoughts.

Social Comparison Theory

Theoretical views. Leon Festinger, a prominent social psychologist, developed a theory stating that people indirectly assess their standing by comparing themselves to others, especially when uncertain about their opinions or abilities (Festinger, 2003;

Taylor et al., 2000). Comparisons are made when objective and concrete basis for judgments are unavailable, leaving subjective assessments as the bases for truth (Festinger). Named social comparison theory (SCT), it states that people strive to achieve accurate assessments of their abilities, opinions, emotions, personality, and acceptability, whether attained intentionally or automatically (Goethals, 1986; Taylor et al.). Judgments are made when people appraise a situation and compare it to what they are capable of doing, leading to future behaviors (Festinger).

SCT states that people's judgments of self-worth and SE are relative to the reference group to which they are comparing themselves (Taylor et al., 2000). Comparisons with others that discredit one's personal perceptions will lead to reductions in SE and lead the individual to seek environments that are more congruent with their ideals (Cast & Burke, 2002). Therefore, people are likely to compare themselves to others who have similar abilities and opinions, attempting to maintain positive self-perceptions through compatible assessments (Festinger, 2003; Taylor et al.). In addition, individuals often associate with those who view them as they view themselves in order to actively verify their self-images (Cast, Stets, & Burke, 1999). Individuals often expect to perform as well as others who are similar and feel displeased when they do not (Goethals, 1986). Therefore, when discrepancies among group members exist, a pressure towards uniformity arises due to feelings of incompetence (Festinger). Once social conformity has been achieved and comparisons become equivalent, self-competence becomes restored. Hence, the drive for social comparison is to increase one's group belonging and maintain feelings of adequacy (Festinger). For adolescents with LQTS, they are likely not able to compare themselves to peers with the syndrome on a frequent basis. When weighed

against healthy peers, discrepancies will exist and social conformity may be unattainable, reducing their SE. If compared to others with LQTS, a group that has similar characteristics and abilities, conformity would be more likely, increasing feelings of belonging and positive SE.

Upward and downward social comparisons. When making comparisons, it is possible to be weighed against those whose characteristics are superior or worse. Some studies have indicated that making downward comparisons to those less fortunate is self-enhancing, while looking up to others who are more successful serves as an indicator of the need for self-improvement (Taylor et al., 2000). Other literature has stated that upward comparisons can signify that you are not as “well off” as everyone or a signal that improvement is possible, which is self-enhancing (Buunk, Collins, Taylor, Van Yperen, & Dakof, 1990). Likewise, downward comparisons can indicate that one is not as “bad off” as others or can suggest that things can get worse. As an example, an LQTS patient comparing himself or herself to someone more academically talented would be an upward comparison, leading the comparator to either feel inadequate or motivating them towards success. Alternately, being compared to a peer with lower grades would be a downward comparison, either boosting one’s feeling of achievement or alerting the comparator to the risk of failure (Taylor et al.).

Researchers have found mixed results for the above hypotheses. For instance, Buunk et al. (1990) hypothesized that people enhance their SE by making downward comparisons, although findings indicated that downward comparisons made by cancer patients led to negative effects. The researchers hypothesized that these results were due to the subjects’ fears of not recovering and uncertainty about their illness (Buunk et al.).

It is also possible that initial feelings of uncertainty about their illness increased subjects' attention to negative information, leading to further reduced feelings of confidence (Buunk et al.). Alternately, Morse and Gergen (1970) found that job applicants indicated having higher levels of SE when their "competition," who was a confederate of the researchers, was disorganized and messy, as opposed to neat and articulate. Another study found that breast cancer patients preferred information and emotional support from well-adjusted targets, acting as models for self-improvement (Stanton, Revenson, & Tennen, 1999). Interestingly, although preferring upward comparisons, subjects often found a dimension to compare downward, such as remarking "they're in denial of their condition" (Stanton et al.). Hence, reducing the discrepancy between themselves and an upward comparison maintained their feelings of competence. Therefore, the impact of comparisons, whether positive or negative, depends upon the interpretation of the situation and expectations of success (Buunk et al.). For LQTS patients, this theory would state that making downward comparisons to high-risk patients could lead to insecurity about their illness. Alternately, comparing themselves to healthy peers could be motivating or lead to feelings of inferiority.

Factors that impact the effects of comparisons are the likelihood of improving or declining based on the evaluation, motivation levels, SE ratings, similarity to the target, and perceived controllability of one's progress (Buunk et al., 1990; Collins, 1996). For instance, Buunk et al. found that patients who believed they were able to control their symptoms and illness outcomes were less threatened by downward comparisons. Another study found that subjects with high SE responded more favorably to upward comparisons than those with low SE (Collins). Individuals with low SE may engage more in

downward assessments in order to increase their feelings of worth (Goethals, 1986). In an analysis of the literature, Collins also concluded that people who shared distinct attributes with the target were more likely to react positively to upward comparisons, since they expected there to be additional shared positive characteristics. The social comparison literature thus indicates that SE both moderates the associations among social comparisons and outcomes, where preexisting SE leads to more favorable assessments, and points to the notion that comparisons can create and maintain SE. Thus, these findings would indicate that adolescents with LQTS who have preexisting levels of high SE would not be greatly impacted by comparisons to healthy peers and would be less affected by comparisons to high-risk patients. Alternately, those with low SE may be prone to the negative effects of upward comparisons to healthy peers and feel more threatened by comparisons to high-risk LQTS patients. It is therefore critical to advocate for adolescents with LQTS by educating school districts about the syndrome and the impact of social comparisons. It is through increased awareness that appropriate support and handling of LQTS-related issues can be provided.

In order to maintain adequate feelings of SE when consistently outperformed, individuals often deemphasize their closeness to the target, downgrade the relevance of the dimension, or hinder the target's performance (Alicke, LoSchiavo, Zerbst, & Zhang, 1997). For instance, one study found that learning disabled adolescents diminished less favorable comparisons and valued positive comparisons, maintaining positive self-evaluations (Crabtree & Rutland, 2001). In other attempts to maintain SE, people will exaggerate the ability of those who unambiguously outperform them, discounting the initial comparison to make the initial comparison seem unfair (Alicke et al.). One study

found that subjects who intensified the skills of the target rated their intelligence as higher, compared to subjects who did not change their perceptions of the target (Alicke et al.). Thus, even seemingly objective social comparisons are mediated by subjective interpretations. Consequently, youth with LQTS may evaluate the athletic and social competence of their peers as being unmistakably outstanding, diminishing the relevance of comparisons, maintaining positive self-perceptions. Therefore, schools can advocate for the student with LQTS by creating activities that have been medically approved, such as recreational noncompetitive games. Being able to partake in athletics may decrease social comparisons, maintaining their positive SE.

Social comparisons and adolescent self-esteem. SCT also helps to explain the nature of peer evaluations. As Harter (1995) indicated, adolescents' selves become increasingly differentiated, creating more characteristics from which to compare, increasing the chances for negative evaluations. Teens are also cognitively able to make self-reflections, highlighting social discrepancies (Renick & Harter, 1989). For example, one study found that female preadolescents and adolescents increased their comparisons to models in advertisements, especially when they had low SE (Martin & Kennedy, 1993). Additional results indicated that adolescents' self-perceptions of physical attractiveness decreased over time, potentially due to increased upward comparisons of the picturesque models (Martin & Kennedy). Hence, adolescence is a developmental period in which individuals with LQTS are likely to increase their social comparisons to healthy peers. Since the ability to self-reflect and think abstractly improves at this stage, it is likely that differences will become more apparent and reduced SE could result.

Researchers have also concluded that the stronger the attraction to the social group, the greater the pressure is towards uniformity among beliefs (Festinger, 2003). As social groups become more valued in adolescence, the importance of avoiding rejection and maintaining peer status increases (Goethals, 1986). Individuals who are similar in attributes believed to be correlated with group categorization are labeled the in group, while those who are different are the out group (Stets & Burke, 2000). Additionally, power is often ascribed to in-group members, leading others to aspire for group membership (Stets & Burke). For example, one study found that people with higher social status, who were more esteemed, had greater influences on others' judgments (Cast, Stets, & Burke, 1999). Therefore, compliance with those in power is perceived to have reinforcing properties. When group compliance is unattainable, continued comparisons may create feelings of hostility and rejection, which could in turn lead to behaviors that attempt to gain approval (Festinger). For this reason, reduced SE may transpire for adolescents with LQTS who are unable to conform to group norms. To corroborate this statement, one study found that learning disabled adolescents who compared themselves to mainstream students had lower scholastic competence ratings than if compared to other remedial students (Renick & Harter, 1989). In addition, competence ratings in relation to the regular education students decreased over time, indicating that differences in social comparisons become more sensitive throughout adolescence (Renick & Harter). These findings indicate that self-perceptions are relative to the comparison group and the dimensions of salience. Additionally, this research signifies that the tendency towards social acquiescence in adolescence assists in the maintenance of SE by increasing social acceptance and solidifying one's self-evaluations.

For youth with LQTS, activity restrictions, medical regimens, and school-based accommodations may highlight their differences, categorizing them as the out group. Additionally, since compliance to group norms is unattainable or could lead to further symptoms, continued comparisons may lead to feelings of inadequacy or failure.

Much of the SCT literature reviewed is associated with one's cognitive attributions, since social comparisons are significant only after being translated into mental interpretations (Goethals, 1986). For instance, maximizing and minimizing perceived similarities are mental processes containing preexisting beliefs, expectancies, and biases about one's abilities (Collins, 1996; Stanton et al., 1999). The SCT literature is therefore relevant to the LQTS population, as personal beliefs about the syndrome, perceptions of symptom susceptibility and severity, individual goals, and definitions of success ascribe meanings to social comparisons, leading to improved or reduced SE. For instance, an adolescent with LQTS who is satisfied watching sports games would have vastly different self-perceptions compared to someone who believes that their social status hinges upon being the star quarterback. Hence, it is critical to understand cognitive processes and their associations with SE.

Cognitive Theory

Theoretical views. At the core of cognitive theory is the belief that individuals' interpretations of events determine how they subsequently feel and behave (Leahy, 2003). Specifically, cognitive theory espouses the theory that subjective thoughts and selective interpretations intervene between environmental stimuli and behavioral and emotional responses (O'Brien & Kollock, 1997). Bandura (1999) stated that thoughts are

creative and reflective; they exert influence on behavior, assess situations, plan for the future, select behavioral options, and evaluate one's adequacy. Additionally, cognitions determine environmental salience, assign meanings, decide emotional impact, and establish motivation (Bandura). Therefore, people's behavior and feelings of self-worth may depend upon mental analysis, whether true or exaggerated, as opposed to objective reality (Taylor et al., 2000).

The attainment of success and self-worth can be conceptualized through the lens of mental models, rules, and predictions of goal attainment (Bandura, 1999). Criteria for competence are based on the values individuals ascribe to them, goals that are set, and standards which they are evaluated against (Bandura). Similar to the symbolic interactionist (SI) view, individuals construct their reality through the interpretations of thoughts, emotions, and behaviors, which influence each other bidirectionally (Bandura; Leahy, 2003). In creating goals, forethought allows individuals to project the likely consequences of their behaviors and assess potential outcomes (Bandura). Predictions are made based on probable environmental contingencies, although SE can become skewed if one's projections are based on uncertainties and ambiguities (Bandura). Lastly, cognitive maturation and development, such as the changes between childhood and adolescence, also contribute to the ways in which perceptions are made (Noppe, 1983). For instance, one study found that adolescents' self-concept was based on concrete ideals for 12-year olds and abstract ideals for 16-year-olds, indicating that age-related changes are associated with different anchors for self-concepts (Noppe). SE is therefore created, evaluated, and assessed through cognitive decision making, which is influenced by development and one's unique world view. For youth with LQTS, this theory would

indicate that feelings of self-competence are based upon the goals that each individual sets and the prediction of the likely consequences. If an adolescent sets a goal to play high school basketball and forecasts that their doctor will allow them to play, their subjective feelings of SE would be based upon the realization or obstruction of that mental expectation. Schools can collaborate with medical professionals in devising plans that will help these adolescents achieve their personal goals, such as modifying gym curricula to allow some level of participation, making reduced SE less likely.

Automatic thoughts. Automatic thoughts are ideas that come spontaneously and appear valid, although they may be misleading, biased, or distorted (Leahy, 2003). If one's immediate thoughts are faulty, they can lead to exaggerated, personalized, or negative interpretations (Leahy). Consequently, stressful and aversive states are often developed, maintained, and strengthened through one's cognitive biases, as opposed to innate environmental causes (Leahy). Automatic standards can also become too rigid or impossible to obtain, impacting subjective self-perceptions and SE (Leahy). For example, if an adolescent with LQTS fails to meet their unrealistic personal social goal of being liked by everyone in their grade, perceptions of rejection may ensue, reducing feelings of SE (Leahy). In this case, the automatic thought of needing to be liked by everyone, as opposed to holding the belief that a few good friends is satisfactory, reduces SE. Rapid assumptions about self-efficacy, which are not analyzed for accuracy, may lead people to discount their abilities and limit opportunities (Bandura, 1999). For instance, an individual with LQTS may hold the automatic thought that they are unlike all other students, thwarting any attempts to make new friends. If analyzed for accuracy, it could be discovered that they have many similar attributes to their peers, such as fashion style,

entertainment interests, and hobbies. Hence, these hasty thoughts can lead to cognitions that ultimately impact SE.

Automatic thoughts also influence behavior when one anticipates the social reactions of others, rather than systematically assessing the likelihood of events (Leahy, 2003). For instance, if an adolescent with LQTS automatically anticipates that others will tease them for receiving school-based accommodations, such as an alternate gym class curriculum, it may influence the likelihood that they will seek out new friendships. Therefore, jumping to assumptions about the likelihood of others' reactions can lead to emotional vulnerability and changes in self-worth. Maintaining the automatic belief that others will react negatively to one's actions may lead to withdrawing from social groups (Leahy). Peer avoidance then creates a vicious negative cycle, since retreating increases feelings of dissimilarity, which then increases additional withdrawal. It is thus important to avoid mind reading and critically analyze one's own assumptions.

In contrast to faulty beliefs, some negative automatic thoughts may be accurate (Leahy, 2003). For instance, an individual with LQTS may believe that their athletic ability is not as strong as other students'. Although this may be a true belief, it can still have a negative impact on SE. Whether accurate or exaggerated, one's thoughts, feelings, and behaviors are connected and interrelated and can be altered with cognitive shifts. Barrett, Webster, and Wallis (1999) found that modifying negative thinking, problem solving, and perspective taking improved SE ratings, self-perceptions, and perceptions of peer relationships among tenth grade students. Therefore, even when one's negative beliefs may be accurate, such as about not being a talented athlete, the increased use of adaptive cognitions can be beneficial, such as recognizing other skills and abilities.

Schemas. Cognitive interpretations are also based on schemas, which are preexisting categories of thought used to selectively attend, to interpret, store, and recall information (O'Brien & Kollock, 1997). Information that is consistent with one's schema will likely be attended to, further reinforcing the initial beliefs, while inconsistencies are typically discounted (Leahy, 2003). For instance, if a middle-school student with LQTS has a negative schema that he or she is are unpopular, and a student tells them not to sit at the lunch table, it is likely he or she will selectively attend to that one incident, discrediting the number of friends they do have. One's automatic thoughts, schemas, and selective filters therefore enhance or diminish SE. The impact of schemas on self-worth often depends on the attributions that one ascribes to the situation.

Cognitive attributions. Attribution theory is a component of cognitive theory, stating that individuals seek out the causes of outcomes, explaining why things work or fail (Santrock, 2001). In essence, we try to understand why people do the things that they do (Taylor et al., 2000). Explanations of causality are inferred either through internal attributions, which are inner characteristics, such as personality, attitude, and ability, or external attributions, which are outer situational causes of behavior, including the environment (Taylor et al.). Thus, individuals may infer that successes and failures are due to ability, effort, and intelligence, or to external causes, such as task difficulty, luck, mood, or help from others (Santrock). Researchers have found that individuals who attribute their positive attributes to the core of their personality and negative characteristics to the periphery tend to report higher global SE (Harter & Monsour, 1992). Some argue that the rules employed in making attributions are socially learned, as

they lead to subsequent perceptions, which reinforce the initial attribution (Gecas & Schwalbe, 1983; Goethals, 1986; Hilt, 2004).

As related to SE, adolescents who attribute social success to internal characteristics, such as having a friendly disposition, as opposed to attributing accomplishments to external forces such as luck, are more likely to experience increases in SE (Santrock, 2001). Accordingly, failures that are attributed to internal causes likely reduce SE, such as if an adolescent with LQTS attributes being picked last for a gym class activity to the syndrome versus random student selection (Santrock). Furthermore, if successes are attributed to stable characteristics, such as personality traits, SE is more likely to increase (Santrock). Alternately, if negative outcomes are credited to stable traits, future failure is often anticipated (Santrock). Additionally, attributions that affect a wide variety of situations, such as being a good friend in all relationships, versus those that are specific, such as being a good friend to one person, have differential impacts on SE (Hilt, 2004). Globally applying positive traits and attributing negative characteristics to circumscribed situations aids in the maintenance of SE (Hilt). Hence, internalization, stability, and globality are dimensions in which attributions vary (Hilt). Overall, if individuals believe they can manage threats, possessing personal efficacy, as opposed to believing they cannot control the environment, it is likely that they will not experience distress (Bandura, 1999). For individuals with LQTS, given the unstable and life-threatening nature of their condition, they may not feel efficacious in managing their syndrome, potentially reducing their SE. Schools can advocate for adolescents with LQTS by providing counseling services that assess and address potential distress while supporting their medical adherence.

In making attributions of others' behaviors, individuals tend to credit internal dispositions as the cause of actions, a common perceptual bias (Hilt, 2004; Taylor et al., 2000). Unfortunately, these assumptions can lead to SE reductions when people assume the worst. For instance, if an individual with LQTS is not invited to a birthday party, the cause will likely be attributed to rejection, rather than a lost invitation. Thus, the causality for successes and failures is subjectively determined, leaving one's attributions open for interpretation (Taylor et al.). Additional attributional biases include the tendency to accept the first attribution, ignoring contradictory evidence, attributing one's behaviors to situational factors, and self-blame (Hilt).

Selective and biased attention to the encoding, interpretation, and retrieval of attributions can also lead to faulty conclusions (Hilt, 2004). Since faulty assumptions often have negative impacts on achievement, motivation, and peer relations, it is critical to probe and identify these negative attributional styles (Hilt). Both prevention and treatment programs can aid in the systematic analysis of one's cognitions to promote SE improvements. For instance, retraining thought patterns and searching for alternate explanations for individuals with LQTS might aid in the recognition of internal characteristics that lead to positive outcomes, such as friendliness, and of external factors that contribute to failures, such as environmental conditions (Hilt). Depending upon one's vantage point, attributions can therefore serve to enhance or reduce one's perception of the self. Future research should continue to investigate whether attribution errors lead to SE reductions or if reduced SE leads to biased cognitions. Although the current manuscript is not investigating cognitive attributions or schemas, if SE is variable in the adolescent LQTS population, this may be an area for future research.

Synthesis of Theories

Based on the literature indicating that adolescent SE is often impacted by the attainment of peer acceptance and school connectedness (Hall-Lande et al., 2007), and peer approval is predicated upon social conformity and fitting in (Santrock, 2001), it begs the question whether teens, especially those with LQTS, who are restricted from social activities and feel dissimilar from others would be prone to having lower self-esteem ratings. For instance, friendship, peer pressure, and role expectations have been found to influence SE during adolescence (Noppe, 1983). One study found that when adolescents answered the question “Who am I?” they responded in interpersonal terms, and their responses indicated the propensity towards social conformity (Noppe). Identity confusion also emerges when conflicts among self-images exist, including who one is versus whom they would like to be (Harter & Monsour, 1992). Given the demands of differentiation in adolescent social and school roles, teens with LQTS, who are urged to adhere to social and school-based limitations, may therefore experience role contradictions and dissonance, potentially leading to SE reductions (Harter & Monsour).

Since having LQTS may prevent or hinder individuals from engaging in valued social activities, lead to feelings of school disengagement, and create feelings of vulnerabilities, the above-noted SE theories could justify why their SE may be impacted. For instance, the symbolic interactionist view could hypothesize that these adolescents would envision how others view them and reflect that they are perceived to be inferior and dissimilar, assigning meaning to their condition. Social comparison theory could point to the notion that upward social comparisons to peers who are able to fully participate in activities would create feelings of inadequacy, indicating they are not as

good as everyone else. Making downward comparisons to others with more significant conditions may be a signal that things could get worse. Cognitive theory could state that schemas associated with unpopularity and automatic thoughts indicating the need to be a part of all social and school-based activities in order to be liked could lead to withdrawal from social situations, further perpetuating feelings of rejection. Lastly, attributions as to why they are different from their peers could be credited to their internal conditions, as opposed to specific situational factors, leading to feelings of hopelessness.

Research has found multiple relationships between the impact of low SE and risk factors associated with negative self-perceptions, such as emotional and behavioral disorders, suicidal behavior, delinquency, eating disorders, and depression (Harter, 1990). Specifically, adolescent conditions that necessitate social and school-based restrictions, such as certain chronic illnesses, could be associated with critical impacts on adolescents' self-perceptions. Therefore, it would be beneficial for schools to advocate for adolescents with LQTS by aiding in the enhancement of positive SE, decreasing the likelihood of negative outcomes, and reducing the risk factors associated with low SE. A comprehensive school-based accommodation tool and emergency response plan will be introduced to address these issues.

Adolescent Chronic Illness and Self-Esteem

Adolescents and Chronic Illness

Chronic illness definition. Given the unique developmental and maturational challenges that adolescents face, including developing a sense of self and personal competence, chronic illness (CI) can pose stumbling blocks for youth. Although a variety

of descriptions exist, CI is often defined as an ongoing medical condition that lasts for a substantial amount of time, characterized by an unpredictable health course, reduced physical performance, and prolonged dependence on medical specialists (Boice, 1998; Kyngas, Kroll, & Duffy, 2000). The impact of CI often necessitates multiple adaptations, including interpersonal, cognitive, emotional, physical, and behavioral domains (Stanton, Revenson, & Tennen, 2007). For instance, lifestyle changes associated with CIs are hospitalizations, surgeries, scheduled medication regimens and side effects, regular health care visits, fears, grief, school absences, and activity limitations (Boice; Woodgate, 1998). Since most healthy children do not encounter these experiences, adolescents with CIs may find it more challenging to become accepted by peers and conform to the teen culture (Boice). Overall, these life alterations have been found to interfere with adolescent peer relationships, role differentiation, and the ability to connect with one's school culture (Boice; Woodgate). To elucidate this point, Vitulano (2003) stated that making friends, maintaining adequate SE and school performance, and participating in sports activities are the most significant issues affecting children and adolescents with CIs as the attainment of these milestones may be limited due to medical regimens and treatment protocols, leading adolescents to feel inadequate and damaged (Vitulano).

Impact of chronic illness on adolescents. The literature has indicated that CIs can impact adolescents in numerous psychosocial domains, including emotional well-being and behavioral, psychological, and social adjustment (Cadman, Boyle, Szatmari, & Offord, 1987). One study indicated that adolescents with a CI are at the highest risk for psychological disturbance (Jarman & Oberklaid, 1990). In a time when acceptance and peer comparison are pivotal, adolescents with CIs may feel more removed from their

social groups. For instance, Blum (1992) stated that adolescents with CIs tend to be more depressed, as they often feel more socially isolated due to restrictions or stigma. Another study found that children with a variety of CIs were at three times greater risk for psychiatric and social adjustment problems and had fewer friends, compared to healthy peers (Cadman et al.). In a qualitative study, Woodgate (1998) found that adolescents experienced extra effort, including physical, mental, and emotional energy, pain, restrictions, and worries, due to their CI. Overall, subjects' themes in this study indicated that "it's hard" living with a CI (Woodgate). Specifically, adolescents highlighted that life becomes more difficult and obstacles are more frequent, including treatment regimens, controlling their symptoms, and incorporating their disorder into their daily lives (Woodgate). Furthermore, participants indicated experiencing "pain" from the restrictive nature of the illness, including limits on social activities, feeling unaccepted by friends, and sensing that they are different from others (Woodgate). In this study, living with a CI increased feelings of frustration, anger, guilt, and hopelessness among adolescents (Woodgate).

Certain illness characteristics can also increase risks for psychological distress, such as prognosis uncertainty, chronic pain, and symptom invisibility (Huure & Aro, 2002). Boice (1998) stated that the type and degree of physical impairment, illness visibility, uncertainty of syndrome course, and unpredictable symptoms are the five CI factors related to psychological outcomes. Researchers have also made multiple hypotheses about the correlation between CI and self-esteem (SE). For instance, it has been argued that poor self-esteem is an antecedent to adjustment problems in youth with CIs (Cadman et al., 1987; Vitulano, 2003). Others have claimed that physical and

psychological stress caused by CIs leads to reductions in SE (Cadman et al.; Vitulano). Hence, given the potential negative effects of CIs and the detrimental consequences of low SE, it is critical to investigate the psychosocial impacts of CIs on adolescents to provide schools with guidelines to maximize school connectedness, safety, and positive SE.

Adolescent Long QT Syndrome and Self-Esteem

Characteristics of Long QT Syndrome

Potential effects of long QT syndrome. As a unique CI, the effects of long QT syndrome (LQTS) may impact adolescents' psychosocial functioning. Although LQTS patients may feel healthy, the syndrome meets the criteria for the definition of a CI, as it an ongoing medical condition, patients' health course is unpredictable, physical performance restrictions are recommended to varying degrees, and patients depend on medical specialists (Boice, 1998; Kyngas et al. 2000). Considering adolescent developmental tasks, such as peer differentiation and advances in self-perception, limitations associated with LQTS may be predictive of SE ratings. In particular, the social and co curricular restrictions the syndrome necessitates to prevent irregular heart rhythms, the school accommodations to maximize education, age of diagnosis, the unpredictability of LQTS symptoms, psychosocial functioning, and potentially compromised school performance may lead adolescents to feel different from their peers. Ultimately, these negative self-perceptions may affect their global self-esteem (SE).

Currently, there are limited studies investigating the psychosocial impact of living with LQTS. Therefore, the hypotheses pertaining to the effects of LQTS need to be

generalized from other cardiac conditions, somewhat comparable CIs, and the general SE literature. Due to unique syndrome characteristics, the LQTS population may be a difficult population to generalize to, since patients face an unpredictable fate, although they are often symptom free. For these reasons, school guidelines, tailored to the needs and strengths of the LQTS community, are in great need.

Generalizations from Other Chronic Illnesses

Other cardiac illnesses and self-esteem. Since there is limited research investigating the impact of LQTS on SE and psychosocial well-being, literature on other cardiac conditions may be informative. For instance, Cohen, Mansoor, Langut, and Lorber (2007) found that adolescent heart disease patients' ratings of social limitations, worries, and frequency of medical examinations were correlated with their perceptions of disease severity, depressed mood, and SE ratings. Researchers noted that subjective disease severity was related to measures of SE, indicating that one's disease perceptions are just as important as objective ratings (Cohen et al.). Salzer-Muhar et al. (2002) found that male adolescents with congenital heart disease had reduced SE and lower self-concept compared to healthy peers. The authors hypothesized that this discrepancy was due to reduced physical ability, which in turn interfered with subjects' peer relationships (Salzer-Muhar et al.). Specifically, boys with heart disease may be especially prone to feelings of ostracism and failure, since their peer group activities surround athletics, while their condition mandates physical restrictions (Salzer-Muhar et al.). In another study, parents rated their children, who had a diagnosis of heart disease, as having poor school results, more school problems and school retention, social and attention problems,

peer teasing, and aggressive behavior compared to a healthy age-matched control group (Miatton, DeWolf, Francois, Thiery, & Vingerhoets, 2007). In the same study, children also reported more depressive symptoms than the control group (Miatton et al.). Once again, the authors indicated that restrictions from sport and social activities hinders social acceptance, potentially contributing to the study's results (Miatton et al.). Furthermore, wanting to participate, but not being accepted by others in social settings may contribute to sadness (Miatton et al.). Although adolescents with LQTS and heart disease may experience different illness trajectories, their social experiences and self-perceptions may be similar.

Other chronic illnesses and self-esteem. Literature pertaining to other adolescent CIs may also generalize to LQTS patients. For instance, cystic fibrosis (CF), a progressive genetic disorder without a cure, characterized by reduced pulmonary function, often sets adolescents apart from their peers physically and socially, due to the athletic and activity restrictions (Christian & D'Auria, 2006). Although CF requires frequent hospitalizations, it parallels LQTS in that patients are barred from athletic activities. In their study, D'Auria, Christian, Henderson, and Haynes (2000) found that school absences, even when brief, interfered with CF patients' ability to participate in school and increased their feelings of displacement upon return. In another study, adolescents with CF indicated that overt behaviors associated with their condition, such as taking medications and leaving activities, highlighted their differences from their peers (Christian & D'Auria, 1997). Another study found that a social skills intervention designed to teach children with CF about their disorder, help adolescents deal with teasing, and find ways to "keep up with their peers" decreased the perceived impact of

the illness and improved global self-competence and self-worth (Christian & D'Auria, 2006). Furthermore, finding good friends who accepted their disease helped to bring a sense of normalcy, support, and approval into adolescents' lives (Christian & D'Auria, 1997). Hence, connecting with peers was found to improve self-perceptions among CF patients, a result that may generalize to the LQTS population. Educating schools and medical professionals about the importance of peer involvement is a powerful advocacy tool.

Epilepsy, a neurological disorder affecting adolescents, is characterized by the occurrence of intermittent seizures (Marin, 2005). Similar to LQTS, epilepsy poses obstacles to youths' independence due to medication management, perceived stigmatization, and an unpredictable symptom course (Marin). For instance, Marin found that adolescents with epilepsy had increased ratings of depression due to unpredictable illness course, perceived lack of control, uncontrollable medical events, and negative self-perceptions. Similar to the cystic fibrosis findings, this study also indicated that taking medications was perceived as a sign of being different and a constant illness reminder (Marin). Additionally, adolescent patients were cautious about disclosing their disorder to peers for fear of rejection and stigmatization (Marin). Similar to LQTS, epilepsy symptoms are often unpredictable, and the fear of having episodes in front of others is ever present (Marin). Thus, Marin stated that interventions, such as school accommodations and promoting medication management may decrease perceived lack of control and minimize the risk for public symptoms, increasing one's self-perceptions.

Research on adolescent diabetes, a CI characterized by irregular blood glucose levels, also points to the impact of activity restrictions on adolescent psychosocial

functioning (Helgeson, Snyder, Escobar, Siminerio, & Becker, 2007). For instance, in their study, Helgeson et al. found that adolescents with diabetes reported less social acceptance compared to healthy peers. Over time, females also began to report increases in depressive symptoms and anxiety, and decreases in self-worth (Helgeson et al.). This study also pointed to the notion that adolescents' yearning to be involved in extracurricular activities and conform to social pressures may have lead to decreased medication management, increasing their symptoms and reducing self-perceptions (Helgeson et al.). A double-edged sword therefore emerges in which taking medications creates feelings of difference, leading to noncompliance, which ultimately increases symptoms and feelings of dissimilarity.

In their study, Helgeson and Novak (2007) interviewed adolescents with type 1 diabetes before and after their routine clinic appointments. Results indicated that females who defined themselves in terms of their illness and viewed it in negative terms had increased depressive symptoms, anxiety, and anger and lower SE (Helgeson & Novak). Hence, when a CI is rated as a central component of oneself, SE may be impacted (Helgeson & Novak). Alternately, when an illness is not considered a central self-component, illness setbacks, such as increased symptoms, may not be as meaningful, although medication adherence may decrease as a result of illness detachment (Helgeson & Novak). This article also points to the importance of the attributions one ascribes to an illness, whether positive or negative. SE interventions should therefore address cognitions associated with illness characteristics in order to maintain positive self-perceptions.

Another chronic health condition that may parallel LQTS, in terms of symptom unpredictability and the potential for fatality, is severe food allergies. For instance,

Avery, King, Knight, and Hourihane (2003) found that children with peanut allergies felt more restricted by their limitations, had higher rates of anxiety, and worried about being away from home compared to children with insulin-dependent diabetes. Compared to the control group, children with peanut allergies were more aware of the potential fatal nature of their condition, leading to heightened anxiety (Avery et al.). Therefore, LQTS patients may also experience concern about potential cardiac episodes, possibly leading to reduced feelings of self-competence. Although the above-noted conditions have similar characteristics to LQTS, its unique disease characteristics may preclude generalizability. Thus, it is important to consider the unique challenges that LQTS poses for adolescents when considering the SE literature.

Challenges to Adolescent Development Specific to Long QT Syndrome

Social restrictions and psychosocial functioning. Christian and D'Auria (2006) stated that psychosocial adjustment is impacted by children's ability to perform developmentally appropriate social and physical activities with peers, especially during middle adolescence. Research has indicated that CIs can impact adolescents' social functioning and SE, since lifestyle changes and adaptations are often necessitated (Kyngas et al., 2000). One study found that SE is derived from the degree of restriction from individual autonomy, the amount of individual control, and the ability to produce intended outcomes (Gecas & Schwalbe, 1983). For instance, a qualitative study investigating adolescent CI found that teens perceived their social limits to be restricting, viewing them as a hardship that prevented them from leading a normal life (Woodgate, 1998).

Since adolescents' identity development is largely dependent upon their active involvement with peers, social restrictions may ultimately hinder peer acceptance, reduce SE ratings, and increase social isolation (Boice, 1998; D'Auria et al., 2000). If one's behaviors do not allow for social identification with a group, feelings of loneliness may also follow (Stets & Burke, 2000). For instance, individuals who are actively excluded from a group may feel depressed, withdraw from others, and increase their dependence on their parents, hindering differentiation and peer conformity (Jarman & Oberklaid, 1990). Frequent peer victimization, including physical and verbal maltreatment, has been associated with poor academic functioning, as mediated by depressive symptoms, including impaired concentration, loss of energy, dysphoria, and negative self-concept (Schwartz, Gorman, Nakamoto, & Toblin, 2005). In their study, Juvonen et al. (2000) found that attributions mediated the relationship between peer victimization and psychological difficulties. Hence, self-blame after harassment lead to psychological maladjustment and later school difficulties (Juvonen et al.). Therefore, the impact of social isolation may be magnified by subsequent reductions in psychological well-being, impacting SE. Alternately, friends and social supports can buffer the effects of CI-related stress and illness flare-ups, leading to greater school success and enjoyment (Vitulano, 2003).

Social restrictions associated with LQTS, such as on sleep overs, sports-related events, and concerts, may therefore impact patients' self-esteem. Since in-group inclusion enhances SE and LQTS restrictions may prevent individuals from engaging in customary activities, these youth may not be as readily accepted (Stets & Burke, 2000). Given that group conformity may be harder to accomplish, social withdrawal, reduced social

identity, and lower SE may result (Kyngas et al., 2000; Stets & Burke). Furthermore, social comparisons to others who are healthy and able to fully participate may lead youth to view themselves as less competent, reducing their self-perceptions. Consequently, at a time when social development is critical to achieving positive self-perceptions, LQTS patients may need to find alternative avenues to connect with peers, which schools can provide guidance with. It is therefore important for adolescents with LQTS to receive support and advocacy through their schools, a place in which social and emotional development occurs.

Co curricular restrictions. School-based, structured extracurricular school activities are a place for adolescents to explore their identity, discover social networks, develop skills, and form trusting relationships (Feldman & Matjasko, 2005). Participation and success at activities valued by peers, such as sports, are a means for adolescents to gain status and acceptance (Donaldson & Ronan, 2006). Positive adolescent development outcomes, including higher academic performance, reduced dropout rates, lower substance abuse rates, less sexual activity, higher SE, less anxiety, reduced feelings of social isolation, and less delinquent behavior, have been positively correlated with school activity involvement (Daniels & Leaper, 2006; Feldman & Matjasko). Restrictions from participation in youth-related activities can therefore lead to social isolation, rejection, and victimization (Poulsen, Ziviani, Cuskelly, & Smith, 2007). Extracurricular activities have also been hypothesized to promote developmentally appropriate prosocial behavior, protect risky behavior, and link adolescents to positive role models (Feldman & Matjasko). Feldman and Matjasko stated that involvement in after-school activities impacts adolescents' psychological well-being by validating their talents, thus increasing

their SE. Likewise, restrictions from participating in co curricular activities may therefore lead adolescents with CIs, including those with LQTS, to feel like outcasts.

Sports participation, in particular, is often considered a rite of passage for adolescents, providing opportunities for peer affiliation and identity formation (Vitulano, 2003). For adolescents with restrictive CIs, this form of social pride may be a struggle to achieve (Vitulano). Competitive athletes are often regarded as heroes and the healthiest group in society (Corrado, Basso, & Thiene, 2005). Male athletes have been found to have high levels of athletic competence, which carries over to their psychological development and their perceptions of self-worth (Todd & Kent, 2003). Craft, Pfeiffer, and Pivarnik (2003) found that females' global self-worth was also correlated with peer acceptance, perceived physical appearance, and perceived athletic abilities. Furthermore, researchers found that peer acceptance was the best predictor of ratings of athletic competence, indicating a relationship between approval and physical self-worth (Craft et al.). Bowker (2006) concluded that physical SE, including perceived physical appearance and physical competence, mediated the relationship between sports participation and global SE. Therefore, for those who do not have confidence in their athletic abilities, lowered global SE may result, while participating in sports may aid in the maintenance of positive SE by increasing feelings of physical competence. Based on these findings, it could also be hypothesized that athletes who must cease sports participation after LQTS was diagnosed may be at risk for reductions in feelings of competence and self-worth, diminished social acceptance, and shifts in their personal identity. Hence, gym class accommodations, such as allowing the student to coach or referee games, can maximize sport involvement while minimizing health-related risks.

Additional studies have investigated the association between increases in sports participation and SE. For instance, in their study, Donaldson and Ronan (2006) found that, among young adolescents, increased time spent in formal sports participation was positively correlated with emotional and behavioral well-being, including less externalizing and social problems and increased athletic, social, physical, behavioral, and global self-worth. Furthermore, self-reported perceptions of sports competence were negatively correlated with internalization, withdrawal problems, somatic complaints, anxiety, depression, social problems, and attention problems (Donaldson & Ronan). Similarly, in their longitudinal investigation of adolescent physical activity level, Stein, Fisher, Berkey, and Colditz (2007) found that increases in physical activity were associated with elevated social and athletic competence scores, while decreased activity showed declined competence ratings. Kirkcaldy, Shephard, and Siefen (2002) also found associations among participation in endurance sports and physical health, psychological well-being, lower anxiety, and depression. Therefore, organized sports participation may enhance adolescents' self-concept and help them to gain overall confidence and social skills by providing more opportunities for social interactions (Donaldson & Ronan; Kirkcaldy et al.). Furthermore, the physical benefits that exercise provides, such as reducing body mass, may also lead to positive social feedback and peer group acceptance, leading to more favorable self-perceptions (Kirkcaldy et al.).

Although research focuses primarily on the benefits of sports participation, one might also consider potential downfalls, such as pressures regarding competition, rejection due to lack of coordination, or embarrassment if one does not make the team. For instance, in their study, Schmalz and Kerstetter (2006) found that sports were

associated with gender stereotypes, and if the males strayed from norms by participating in “feminine” athletics, their masculinity was questioned. Alternately, girls who did not participate in “feminine” activities, including dance, ballet, or gymnastics, experienced stigma consciousness, feeling self-conscious about their gender identity (Schmalz & Kerstetter). Therefore, although research indicates that there are benefits to engaging in structured athletics, there are also potential costs and downsides.

In thinking about LQTS patients, who may be restricted from sports activities in order to prevent heartbeat irregularity, their self-perceptions and ultimate SE may therefore be compromised. Being excluded from organized competitive sports may make individuals feel like outsiders, providing them less social opportunities. Additionally, not conforming to other peers by partaking in games may set them apart from other students as “different.” For instance, in a study investigating participation in team sports among adolescent males with coordination disorders, Poulsen, Ziviani, Cuskelly, and Smith (2007) found that amount of time spent participating in team athletics mediated loneliness ratings, regardless of level of physical coordination ability. Hence, increases in sports participation were associated with decreased loneliness (Poulsen et al.). It was hypothesized that highly structured peer groups provided affiliation, supportive associations, emotional involvement, leadership, self-efficacy, and behavioral competencies (Poulsen et al.).

As a potential resolution to activity restrictions, Vitulano (2003) recommended that children be offered alternative roles, such as manager, statistician, or assistant coach. Although adolescents may not have an active role as a team participant, they may therefore be able to maintain feelings of belonging by maintaining involvement in

another capacity. Alternately, these special accommodations, in themselves, may lead to feelings of being singled out, leading to the same outcome. Donaldson and Ronan (2006) indicated that defining athletic success in terms of personal goals might aid in the enhancement of perceived athletic competence, potentially leading to better overall SE. Therefore, sports ability should not be determined by making social comparisons, but through the assessment of individual abilities. Lastly, Poulsen et al. suggested that participating in nonphysical activities, such as band, choir, or youth groups, may provide children the opportunity to engage socially with peers, although those activities are often lower in social status. Therefore, there is a need for schools to recognize both the benefits and potential downfalls of special accommodations in order to provide adolescents with LQTS the most successful accommodations.

School accommodations. Adolescents with CIs, such as asthma, cancer, allergies, and immune disorders, often receive educationally related services, including school-based accommodations to maximize academic achievement and accommodate individualized needs (Betz, 2001). The Individuals with Disabilities Education Act (IDEA) entitles eligible disabled students, ages 3 to 21, to free and appropriate education, providing suitable educational opportunities to those who meet the criteria for a disability (Sullivan, Lantz, & Zirkel, 2000). To prohibit disability discrimination, allowing students to attend school and participate as fully as possible, Section 504 of the Rehabilitation Act of 1973, a branch of the IDEA, entitles disabled students, both general and special education students, to accommodations in all federally funded agencies, including public schools (Betz; Sullivan et al.). Under this law, “individuals with a disability” are those who have a physical or mental impairment that limits at least one major life activity

(Betz). Furthermore, individuals must also have a record of the disability and be regarded as having the impairment (Betz). Categories of accommodations provided by Section 504 are learning supports, disease management, and ongoing monitoring, such as adjusting class schedules, providing extended time on tests, administering medications, offering rest periods, assisting with reentry from hospitalizations, managing symptoms, providing staff disease education, and monitoring by teachers (Betz).

As the developmental literature indicates, a prominent social context for adolescent development is the school setting, as it provides youth with academic opportunities and is a central hub for social activities (Hall-Lande et al., 2007). School connectedness has also been associated with protective factors, such as lower levels of psychological problems and violent behavior (Hall-Lande et al.). For youth, SE is often be derived from the self-worth and satisfaction derived from school-based accomplishments (DuBois et al., 1998).

Although intuitively it would appear as though school-based accommodations would improve school connectedness and social acceptance, these modifications are not always welcomed. For instance, adolescents may resist disclosing their conditions to their teachers and school staff, fearing that they will become stigmatized or labeled as different or their condition will be misunderstood (Betz, 2001; Suris, Michaud, & Viner, 2004). Becoming labeled with a disease may also become internalized, affecting a person's identity and self-conceptions (Russell, 1984). For instance, due to the relative rarity of LQTS, there may be a lack of understanding of the syndrome, which could increase levels of adolescent frustration and feelings of difference. Adolescents with LQTS may appear to be perfectly healthy, therefore creating questions surrounding why they receive

differential treatment. Christian and D'Auria (1997) also found that adolescents with cystic fibrosis would often keep secrets and hide dissimilarities in order to reduce differences from peers and avoid getting picked on or gossiped about. Parents may also resist having their children treated differently from others (Betz). In their qualitative study of young people's views of their CI, Lightfoot, Wright, and Sloper (1998) found that teacher reactions to illness and relationships with peers were related to how subjects felt in school. Difficulties emerged when school staff or pupils lacked knowledge or misunderstandings arose, leading some to want their privacy (Lightfoot & Sloper). Unfortunately, keeping critical medical information private can lead to medical emergencies within the school, creating additional feelings of embarrassment and dissimilarity (Suris et al.). Lightfoot, Wright, and Sloper recommended that, as a way to dispel myths about student disabilities, information be provided to school staff about a child's abilities, in an attempt to increase inclusion in appropriate activities.

Research also indicates that adolescents may not derive as much self-satisfaction from their academic accomplishments when they have external aids or supports (Bandura, 1998). For instance, LQTS students who win a game of basketball may not take as much pride in the accomplishment if they were allowed additional breaks or time-outs. Alternately, increases in SE are more likely to arise when success is ascribed to one's own abilities and efforts (Bandura). Even when successes are due to internal characteristics, individuals with accommodations may attribute their achievements to their supports (Hilt, 2004). These attributions could reduce motivation and perpetuate the views that their efforts are fruitless (Hilt). For instance, Christian and D'Auria (1997) found that adolescents with cystic fibrosis felt that unequal standards were used to

evaluate their performance, feeling that coaches and teachers treated them differentially upon learning of their diagnosis. Therefore, whether successful or not, these individuals alleged that they were not treated the same, which impacted their school-perceptions.

School-based accommodations may also increase visible dissimilarities among youth with CIs. For example, in one study, subjects indicated that leaving class to take their medications with the school nurse signified their differences from peers, as students would question their behaviors (Christian & D'Auria, 1997). These experiences may generalize to LQTS patients. Trips to the nurse's office to take medications, gym class accommodations such as breaks from physical activity, teachers' awareness of LQTS, school staff trainings, and emergency school plans may increase perceptions that one does not fit in with peers, ultimately impacting global SE. Given the potential negative impact of school-based services on adolescents with chronic illness, intervention plans should be developmentally appropriate, involve the collaboration of the students and their family, and should be revised as needed.

Age of diagnosis. Adaptations to CI may be influenced by the individual's age at which the condition was first diagnosed. It could be argued that diagnosis at a younger age may be associated with lower SE ratings. As one possible explanation, social support for CI changes over time, potentially leaving adolescents in whom the CI has been diagnosed for a long time with less assistance (Stanton et al., 2007). As research has indicated, adjustment to illness is often correlated with help from others, so reductions in support may also decrease feelings of self-worth and SE (Stanton et al.). Early diagnosis may have also necessitated social restrictions as a youth, so that adolescents are faced with the task of catching up to fit in (Blum, 1992). This, in turn, may lead to greater SE

reductions. Furthermore, misunderstanding about the chronic nature of certain illnesses can lead to increases in relationship strain over time (Stanton et al.). As an example, friends may not understand why individuals with LQTS have not gotten better, becoming more frustrated and intolerant of their limitations. Furthermore, prior illness-related experiences influence how adolescents view their CI course and what they remember and understand about present events (Christian & D'Auria, 1997). If an individual had negative experiences in the past, they will likely reflect negatively upon their illness (Christian & D'Auria). Thus, it is possible that the longer one has had the illness, the greater the chance for increased symptoms, surgeries, or medication trials, leading to lower illness perceptions and self-ratings.

Alternately, a case could be made that recent diagnoses, during adolescence, may be associated with confusion and reduced self-perceptions. For instance, cognitions about one's illness may be impacted by stages of cognitive development (Woodgate, 1998). Adolescents in whom the condition is newly diagnosed may not recognize that living with their CI may become better, blind to the waxing and waning of their condition (Woodgate). Although it is a stage in which abstract thought is emerging, some adolescents may not be able to self-reflect, look into their future, and hypothesize about positive outcomes (Harter, 1995). In contrast, children in the concrete operational stage may not conceptualize their CI as being a long-lasting, integral part of their being (Boice, 1998). Although a potentially inaccurate view, this innocent outlook may serve to maintain positive SE (Boice). For instance, one study found that patients with cystic fibrosis began to understand the importance of medical treatment and became aware of their illness between ages 6 and 8, when first confronted with peer differences at school

(D'Auria et al., 2000). Therefore, during early childhood, they did not understand the implications of their CI. Alternately, having the condition diagnosed in adolescence may create identity confusion, as teens are more self-aware and self-conscious. Hence, if the diagnosis is made at a younger age, it may be easier for adolescents to incorporate their existing self-perceptions into their emerging selves, as opposed to doing both simultaneously. Diagnosis during youth may be associated with higher SE because “normal” would be different for them, as they would only know one way of living, rather than having changes and restrictions that are newly introduced. For example, if the condition is diagnosed in adolescence, individuals who had been athletic would need to stop most activities. Therefore, in addition to making sense of emerging roles and pubertal changes, these adolescents would have to make additional identity shifts, redefining themselves as not eligible to play competitive sports, shifting their personal and social goals.

Frequency of medical events. Frequency of medical events may alter cognitions of vulnerability in LQTS patients, potentially reducing their SE. Disease severity, prognosis, health declines, and symptom presence are correlated with adaptation to one's CI (Stanton et al., 2007). Cognitive appraisals and attributions of threats to health, life, and goals and illness-related expectancies can impact one's adjustment and coping (Stanton et al.). Decreased coping often leads to a reduction in information seeking, problem solving, social support, and optimistic beliefs, which may impact SE (Stanton et al.). Therefore, increases in illness events may decrease confidence in one's ability to effect desired outcomes, leading to thinking patterns of dissimilarity and rejection (Stanton et al.). Alternately, remaining symptom-free can aid individuals in distancing themselves from

their condition, feeling more healthy and in charge of their condition, maintaining positive SE (Stanton et al.).

Frequent medical events may also impact patients' perceptions of illness control. Researchers have found that difficult life events that are out of one's control, such as unpredictable LQTS symptoms, put SE at risk (Baldwin & Hoffmann, 2002). Helgeson (1992) stated that perceived control over realistic aspects of one's illness is an adaptive function, whereas feelings of vulnerability can emerge if one does not perceive power over future events. Findings have indicated that internal feelings of control are associated with better adjustment to illness for cardiac patients (Helgeson). Ross and Broh (2000) found that people who do not feel that they are in control of life outcomes are less persistent in coping, thus leading to less successful problem-solving strategies.

Adolescents' SE may also be related to health behaviors, which impact symptom presentations. Although the teenage years bring increased responsibility for the self-management of CIs, adhering to medical advice often conflicts with developmental tasks, such as conforming to social norms (Williams, Holmbeck, & Greenley, 2002). Medical regimens that may lead to peer exclusion are less likely to be adhered to (Christian & D'Auria, 1997). Consequently, the more medical advice interferes with adolescents' normal lifestyles, the more likely noncompliance will be, increasing symptom frequencies (Kyngas et al., 2000). For instance, adolescence is often a time for experimentation with alcohol, signifying fitting in and being in the in crowd. Since individuals with LQTS are advised to avoid alcohol, they may be less likely to comply, given the conflict with the social norm.

Torres, Fernandez, and Maceira (1995) stated that individuals with higher SE appropriately perceive the consequences of their health-related behaviors and have greater self-efficacy over their positive health habits. Varying health beliefs also determine involvement in health-related activities and medication adherence (Torres et al.). The authors found that adolescent SE was correlated with general health behavior and the value of health (Torres et al.). Specifically, for young adolescents, SE was related to personal health, mental well-being, and health behavior, and SE in older adolescents was related to mental health and health-related safety behaviors (Torres et al.). Hence, SE impacts adolescent health behaviors, which may impact future ratings of SE. Higher SE has also been correlated with following healthcare recommendations, which may reduce medical events associated with CI (Torres et al.). Therefore, more frequent medical events may occur due to disregarding of medical advice, as mediated by SE levels, and low SE may lead to poorer coping, which further reduces treatment adherence. Preventing LQTS cardiac symptoms could therefore increase SE by thwarting medical events.

Lastly, research has indicated that symptom trajectories may influence the meaning adolescents ascribe to their CI (Woodgate, 1998). For instance, one study found that the difficulties associated with CIs were related to the phase of the condition, whether stable or unstable, as opposed to the type of illness (Woodgate). When in remission, it was easier for subjects to cease thinking about their CI, defining their lives as less difficult (Woodgate). In this study, individuals also indicated that worries about health consequences added to the difficulties of living with a CI, increasing their feelings of hopelessness (Woodgate). Therefore, one's CI may take center stage when events are

more frequent, potentially altering self-perceptions. Jarman and Oberklaid (1990) also indicated that when medical conditions are perceived as more pervasive, they lead to greater psychosocial difficulties. Alternately, if individuals integrate their CI into their self-concepts, they may increase self-care, leading to increased health and decreased symptoms (Helgeson & Novak, 2007). Therefore, subjective experiences and physical states impact adjustment. Given these results, increased LQTS medical events, such as syncope or fainting, may intensify feelings of concern, leading to reduced SE.

Treatment adherence may also be a concern for LQTS patients, as they often feel otherwise healthy. As such, adolescents may question the necessity of their treatments and opt to maintain “normalcy.” In addition, younger adolescents may not have the ability to think abstractly into the future, unable to imagine prospective consequences of medical non adherence, leading to reduced compliance (Suris et al., 2004). Therefore, lack of adherence may lead to more frequent symptoms, increasing feelings of unpredictably, reducing SE.

School and academic functioning. School functioning is one of the most vulnerable aspects of adolescent SE, as peer evaluations often take place in the school setting (Sukumaran et al., 2002). Children with CI often perform less well academically compared to healthy peers (Lightfoot & Sloper, 1998). The effects of CI, such as ill health and fatigue, are associated with higher rates of school absences, placing individuals at risk for academic problems and additional feelings of social isolation (Boice, 1998; Jarman & Oberklaid, 1990). One study found that school absences decreased academic achievement scores and reduced feelings of social competence among adolescents with CI (D’Auria et al., 2000). Poor grades and high absenteeism

rates have also been found to increase feelings of loneliness, low SE, and depression (Juvonen et al., 2000). Vitulano (2003) stated that the feelings of social isolation and incompetence are related to suboptimal performance levels and less school enjoyment. Alternately, ratings of high school belonging, including school connectedness and perceived school membership, have been found to predict academic performance and psychological adjustment (Pittman & Richmond, 2007). Hence, school absenteeism and feeling different from one's peers can cause detachment from school, leading to lower academic attainment. As a way for schools to aid students in their return to school, reintegration programs are often implemented, including preparing one's classmates for the return, providing extra sets of books, and allowing students to ease back into their regular schedule. Direct guidelines for LQTS do not exist in any great detail, and therefore this project will be one of the first to provide a comprehensive school-based accommodation tool and emergency response plan, including guidelines for school reintegration.

Academic performance has also been associated with adolescents' ratings of SE. In the SE model of academic functioning, adolescents with higher ratings of personal worth tend to do better in school, which increases feelings of control and high SE, impacting future academic success (Ross & Broh). Filozof et al. (1998) also found that academic performance influenced academic and general SE among high school students, pointing to the importance of academic success in adolescents. Another study found that seventh grade Portuguese students who had lower levels of academic achievement had less favorable attitudes towards school, lower grades, and lower SE (Alves-Martins, Peixoto, Gouveia-Pereira, Amaral, & Pedro, 2002). Furthermore, Portuguese adolescents

with lower academic achievement levels attributed less importance to school, a possible attempt to protect their SE (Alves-Martins et al.). For these reasons, if CI is associated with poorer academic functioning, which impacts SE, it is critical to advocate for the students and aid individuals in maintaining their school success.

Academic attributions, judgments about the causes of school success and failure, also impact achievement. For instance, individuals with high motivation towards achievement often believe their success is due to internal ability and effort (Weiner, 1998). High motivators often initiate activities, work with great intensity, persist, and choose challenging tasks (Weiner). Furthermore, high achievers feel more pride in successful accomplishments, anticipate future success, and recognize the correlation between hard work and success, which reinforces achievement-oriented behaviors (Weiner). Broh (2000) found that feeling in control of outcomes, viewing outcomes as contingent upon effort, and believing success is a consequence of actions, were associated with higher grades and test scores among high school students. In contrast, those low in achievement motivation believe that failure is due to a lack of ability, have lower frustration tolerance, and anticipate future failure (Weiner). Another longitudinal study concluded that middle school children's internal ability attributions, mastery-oriented goals, and persistence predicted perceptions of academic competence and actual academic achievement outcomes one year later (Obach, 2003). For those with unpredictable CIs, including LQTS, it is possible that they may think that outcomes are determined by outside forces (Ross & Broh). Since sporadic symptoms are a hallmark of LQTS, it is possible that adolescents may believe that they are not in control of their health outcomes. In turn, these beliefs of perceived powerlessness over desired results

may lead to lessened efforts in other realms, such as academics. For instance, LQTS patients may hold general schemas and automatic thoughts that goal achievement is dependent upon luck and assistance from others, as opposed to internal characteristics.

The Current Manuscript

Generalizations to Long QT Syndrome

Based on the self-esteem (SE), chronic illness (CI), and adolescent development literature, coupled with theories of symbolic interactionism, social comparison, and cognitive attributions, it is possible that self-esteem problems exist in the adolescent LQTS population. Considering the above-noted literature, adolescent milestones, such as establishing friendships, engaging in athletics, and academic achievement may be harder to achieve for individuals with LQTS. Although they may feel otherwise healthy and live symptom free, the social and exercise-related limits placed on these adolescents, coupled with their personal disease history, may place them at risk for perceiving themselves differently from their peers, reducing their SE. This manuscript will provide guidelines for advocating for adolescents with LQTS by supplying a comprehensive school-based accommodation tool and emergency response plan. Advice and education for schools on what to ask the families, how to maximize SE opportunities, and how to prepare the school to be a supportive environment are in great need. These procedures are therefore intended to support and enhance the psychosocial functioning of students with LQTS while improving their health and safety.

Needed Research

Currently, the field of psychology is lacking research pertaining to the psychological or social functioning of adolescents with LQTS. This thesis is an arm of a greater program of research purported to be among the first pertaining to the psychosocial functioning, quality of life, and well-being of children and adolescents with LQTS. This manuscript aimed to investigate the psychosocial functioning and self-esteem of adolescents with the syndrome. Given the unpredictability of LQTS, coupled with social restrictions, physical restrictions, and unique disease characteristics, this population could benefit tremendously from support from the psychological community, including guidance in the creation of school-based accommodation plans.

This manuscript will assist medical, school-based, and mental health professionals to identify and advocate for some of the psychological and social needs of youth with LQTS. The introduction of a comprehensive school-based accommodation tool and emergency response plan will help raise awareness pertaining to the needs of the population, provide procedures to advocate for the psychosocial needs of adolescents with LQTS, and educate schools about the strengths and resiliency that LQTS patients possess. Hence, recommendations from this thesis may lead to increased advocacy and education, extend research in the areas of needs assessment, guide future studies, provide school-based protocols, and sensitize practitioners to the implications of living with LQTS. In addition, the empirical study that is under way may lead to future awareness of the impact of LQTS on psychosocial functioning and SE of adolescents with the syndrome.

CHAPTER 3

Hypotheses

*Hypothesis and Research Questions**Research Question*

The purpose of the current manuscript was to examine whether long QT syndrome (LQTS), a chronic and potentially life-threatening genetic cardiac illness, impacts adolescents' psychosocial functioning. Although the study was not able to be conducted within the allotted 13-month time frame due to unforeseen logistical challenges, the study questioned whether the self-esteem (SE) of adolescents with LQTS is impacted by the limitations associated with LQTS, social functioning, and school functioning. Specifically, the study questioned if personal LQTS history, including social restrictions, involvement in co curricular activities, school accommodations, age of diagnosis, frequency of events, social functioning, and school functioning measures are predictive of SE in adolescents with LQTS. Data collection is now under way, and data will be analyzed and reported accordingly.

Hypothesis

It was hypothesized that the unique challenges that LQTS impose on adolescents impact their social functioning, school performance, and SE. Specifically, it was hypothesized that adolescents with higher ratings of self-reported social and school functioning would have higher levels of SE. Additionally, it was hypothesized that social and co curricular restrictions, school accommodations, age of diagnosis, frequency of medical events, social functioning, and school functioning would predict ratings of SE.

Given the hypothesized needs of the population of focus and support, based on previous literature and corroborating theories, recommendations for school personnel and a detailed school-based accommodation plan are provided, along with a crisis response plan.

CHAPTER 4

Methods

After 2 ½ years of involvement in the LQTS program of research under the direction of Dr. Stephanie Felgoise (Felgoise, 2008) and 13 months of attempted data collection, it was determined by the research team that the benefits associated with proceeding with an intervention plan for adolescents with LQTS far outweighed the costs of waiting for data. Although the current study was not able to be conducted within the allotted time frame, due to unforeseen logistical issues, including challenges securing the sample of patients, the study hypothesized that adolescents with higher ratings of self-reported social and school functioning have higher levels of self esteem (SE) and that social and co curricular restrictions, school accommodations, age of diagnosis, medical event frequency, social functioning, and school functioning predict ratings of SE. Often, large university hospitals who treat LQTS patients are busy and the procedures for their institutional review boards, the governing bodies that approve research, are frequently lengthy and stringent. This research team recognized that there is sparse research pertaining to adolescents with LQTS, although it was challenging to enroll participants in the study. It is ironic that a population in need of exploration is difficult to secure.

CHAPTER 5

Results

*Accommodation Plans for Adolescents With Long QT Syndrome (LQTS)**Rationale for LQTS Accommodations*

Assuming that the hypothesis for the current investigation would have been supported, the goal of the current study was to create an ecologically valid school-based intervention plan addressing the LQTS variables this manuscript examined, including adolescent social restrictions, co curricular restrictions, school accommodations, age of diagnosis, frequency of events, social functioning, and academic functioning. Empirical literature was used to guide the plan, including the thoughts, behaviors, and feelings and self-esteem concepts reviewed previously. The following are guidelines for schools to model, including a comprehensive system of what to ask, do, and plan in order to support the self-esteem of adolescents with LQTS and integrate them into school. The objectives of this plan are to address the psychosocial needs of adolescents with LQTS, increase student advocacy, minimize the impact of LQTS at school, increase LQTS awareness, and provide parents, schools, and health professionals with guidelines for school-based services and emergency plans. The interventions provided target the most needed areas as indicated by the research, including extracurricular and school-based activities.

The following school-based plan for adolescents with LQTS was modeled after other interventions and recommendations, including that from the Sudden Arrhythmia Death Syndromes (SADS) Foundation (*SADS works to keep children safe at school*, 2009) and the American Diabetes Association (*Diabetes at school*, 2009). The SADS Foundation website provides recommendations for patients with LQTS and various other

arrhythmia syndromes. For instance, the website provides a template for an individualized school health care plan for youth with LQTS and students with pacemakers or implanted defibrillators (ICD), including general emergency response procedures and a chart to document medical emergency information (*SADS works to keep children safe at school*, 2009). Although not individually tailored to each student's needs, the website provides an informational brochure pertaining to LQTS and athletic restrictions, definitions of school-based services, and a general school accommodation checklist for students with any SADS condition (*SADS works to keep children safe at school*, 2009). The SADS Foundation also provides a letter for parents to take to their child's principal, describing LQTS and general emergency care and sample emergency response plans (*SADS works to keep children safe at school*, 2009). Although useful, these plans do not outline the procedures to initiate and monitor the plans, nor do they explain how to collaborate with schools, advocate for the children, and ensure that the plans are individually tailored and monitored. The American Diabetes Association also provides the basic principles of safe care at school, including staff training, disease understanding, medical emergency plans, designated roles, nurse involvement, and medication management (*Diabetes at school*, 2009), which the current plan has modeled. The following recommendations should always be approved by the child's family, school, and medical professionals, as programs must be tailored to individual's unique strengths and needs.

Rationale for School-Based Accommodations

Importance of school involvement. Schools are a critical context in which psychosocial and psychological development transpire, including for children who have chronic illnesses and health conditions (Drotar, Palermo, & Barry, 2004). Schools have the ability to provide many opportunities for health-related prevention and intervention activities, since students are easy to access, multidisciplinary teams are available, interventions occur in naturalistic environments, and accommodations can be readily assessed, monitored, and adapted (Power & Blom-Hoffman, 2004). Schools also lend themselves to implementing health-related services, as they are highly accessible to families, parents can be readily integrated into health plans, community-based relationships exist, and positive peers and role models are available to reinforce health-promoting messages (Power & Blom-Hoffman). Additionally, school staff, such as nurses, school psychologists, counselors, and administrators, are often available to assist children in overcoming potential obstacles to attaining educational and personal achievement, along with supporting their health and well-being. Hence, schools are a prime environment in which youth with chronic illnesses can receive interventions, including plans for medication management, individualized educational programs that maximize school success, health-related service programs, reintegration programs after illness-related absences, programs for peer support, and health and general well-being monitoring (Drotar et al.).

Challenges schools face. Although schools are an optimal environment for medical prevention and intervention programs, there are challenges that schools face in adequately implementing health plans. The prevalence of youth chronic illness has nearly

doubled in the past few decades, and it is estimated that 10% to 15% of children with chronic illnesses attend school on a regular basis (Brown, 2004; Clay, 2004). Federal legislation mandates that schools address the aspects of students' illnesses that may negatively affect their educational success, making schools responsible for thousands of children with chronic health conditions each year (Clay). Unfortunately, schools often do not have guidance in creating intervention and prevention plans, staff frequently lack medical expertise and emergency training, administrators often solely focus on educational achievement, teachers have demanding and overextended schedules, staff is responsible for large caseloads, and schools do not have enough resources or staff to create comprehensive and collaborative health plans (Clay; Power & Blom-Hoffman, 2004). Also, school-based programs are often fragmented and disconnected other health and medical professionals, community services, and family participation, leading to less effective interventions (Power & Blom-Hoffman). In their study, Telljohann, Price, Dake, and Durgin (2004) found that many schools only have part-time nurses, and children's health needs either are not being met or are handled by nonmedical personnel, such as secretaries, teachers, and administrators. In their survey-based study pertaining to youth asthma, a potentially life-threatening chronic health condition, Hillemeier, Gusic, and Bai (2006) found that nurses in Pennsylvania are not able to meet students' asthma needs because the ratio of nurses to students is too low, medical care is often delegated to nonmedical professionals, and fewer than one quarter of schools provide counseling or psychological support for youth with asthma. This study found that only four of five schools had staff who knew what to do in the event of a severe asthma attack, putting students in significant risk (Hillemeier et al.). This was a surprising statistic, given the

relative frequency of childhood asthma compared to LQTS. The authors also found that medical equipment is not available in many Pennsylvania schools, emergency response plans are not always in place, and nurses may not know that a student has asthma until they come into the office with symptoms (Hillemeier et al.). Lastly, the findings indicated that only one quarter of children with asthma had written health plans at school, and of those, many did not include emergency contact information or recommended procedures in case of an event and lacked collaboration (Hillemeier et al.). It is therefore extremely important for schools and families to have access to literature and education pertaining to the creation and maintenance of effective illness-specific accommodation plans. It is also significant to note the diversity within and between schools, individuals, and medical practices, making it challenging to predict all of the obstacles in creating, implementing, and following through with accommodation plans. Although the following characteristics and procedures are a comprehensive compilation of best practices within the literature and should be tailored to each situation, it does not guarantee student safety. On the other hand, given the potential benefit to youth health and well-being, these plans are critical for the psychosocial functioning, self-esteem, and health of adolescents with LQTS.

Accommodation Plan Characteristics

Biopsychosocial model. There are certain characteristics that school-based accommodations should possess. First, the biopsychosocial model asserts that chronic illness is bidirectionally impacted by social, emotional, physical, psychological, biological, and environmental factors (Carlson, Kubiszyn, & Guli, 2004). School-based intervention plans should be multifaceted, including an emphasis on prevention,

management, and adaptation to illness, behavioral contributors to illness, developmental stages, environmental factors, socioeconomic variables, psychological functioning, and social-emotional development (Brown, 2004). According to Rae and Fournier (2004), interventions should be collaborative, child focused, developmentally appropriate, multidisciplinary, assessment driven, empirically driven, and intervention and prevention oriented. Hence, plans should be individually tailored to the unique needs of the student, family, and the school. An important aspect of an accommodation plan is also student advocacy. As this plan will delineate, educational professionals should be informed of the adolescents' special needs, medical budgets should be requested for purchasing medical devices and services, and partnerships with other disciplines should be requested.

Collaboration and consultation models. Utilizing collaboration and consultation models within schools, incorporating medical and mental health fields in the development of accommodation plans is the gold standard within the literature. According to Power and Parrish (1995), a multidisciplinary team approach, including medical, psychological, and educational experts, is needed to assess and address the multifaceted needs of youth with chronic illnesses. Collaborations among families and health systems, families and schools, and schools and health systems are essential for the effective care of youth with chronic illness (Phelps, 2006). According to Phelps, it is critical that these systems link together in order to advocate for the health and well-being of chronically ill children in a school setting. It is through collaboration that efficacious behavioral, instructional, medical, and psychological interventions can be created and implemented (Power & Parrish). The literature indicates that collaboration improves performance, enhances communication, creates continuity among contexts, increases shared ownership and

commitment, improves implementer's follow-through and buy-in, increases understanding of the complexities of health conditions, and helps pool resources (Sheridan & Cowan, 2004). Steps in establishing and maintaining collaboration include identifying a team, establishing goals, determining leadership and roles, communicating openly, planning strategically, conducting continual assessments, implementing plans, problem solving, and thinking ahead (Power, Shapiro, DuPaul, & Kazak, 2003). For example, teachers can often provide information pertaining to classroom-based functioning, behavioral functioning, and observations and implement strategies, parents can provide expertise pertaining to behavioral, medical, developmental, and social concerns, medical consultants can share expertise about LQTS and the impact on the child's functioning, and students can give their feedback and input on their own strengths and needs. Given the unpredictable nature of LQTS, it is therefore critical that students, parents, teachers, nurses, administrators, counselors, and school psychologists collaborate with one another in order to ensure the successful integration of the student into the school, support their positive peer relationships, implement appropriate curricular accommodations, and recognize their medical needs (Clay, 2004). Collaboration should also include exchanging information pertaining to curriculum modifications, medication regimens, medical equipment staff training programs, and crisis interventions.

Prevention and intervention models. The literature recommends that school-based programs employ multiple prevention and intervention strategies. First, school-based health programs often have primary prevention efforts that increase protective factors and reduce risk factors, secondary prevention efforts that aim to reduce the prevalence and severity of an illness, and tertiary prevention that reduces residual effects of an illness

(Brown, 2004). For instance, LQTS primary prevention efforts include restrictions and adaptations to the physical education curriculum to prevent sudden cardiac arrest, secondary prevention efforts are purchasing automatic external defibrillators (AED) to reduce the effects of an arrest, and tertiary prevention is psychological counseling to re-acclimate the student into school after an illness-related absence. Overall, the primary components of the LQTS accommodation include a multidisciplinary intake assessment for prevention and intervention, collaborative development of a medical and educational health plan, staff training, and a disaster response plan. The following section describes detailed steps in creating and implementing optimal accommodation plans for adolescents with LQTS, according to the literature.

Steps in Creating LQTS Accommodation Plans

Background. Youth with a chronic illness that limits one or more major life activities, such as breathing, learning, eating, and walking, are eligible for school-based services (Clay, 2004). The goals of these accommodations are to enable “disabled” students to participate in or benefit from school activities, providing them with equal opportunities within the school setting (Phelps, 2006). Since LQTS is a chronic health condition that places patients at increased for irregular heart rhythms and sudden cardiac arrest, individuals should be eligible for a school-based health plan to accommodate their medical and school-based needs (Napolitano, Bloise, & Priori, 2006).

In June 1997, President William Clinton signed the Individuals with Disabilities Act Amendment (IDEA), which mandate that federally funded schools provide free and appropriate public education (FAPE) to all qualified disabled students (Brady, 2004).

Providing FAPE is critical to schools, as adhering to the amendment allows districts to continue to receive federal funding (Brady). Therefore, under IDEA, schools receive federal funding in order to meet the educational needs of students with disabilities (Brady). Eligible IDEA categories include autism, specific learning disability, speech or language impairments, emotional disturbances, traumatic brain injury, visual impairment, deafness, mental retardation, deaf-blindness, multiple disabilities, orthopedic impairment, and other health impairment, which must adversely impact educational performance (Brady). As a part of IDEA, schoolchildren are eligible to receive educationally related services to maximize their school success, such as speech and language, physical therapy, and school health services (Lee & Janik, 2006). Eligible students under IDEA also qualify for individualized educational plans (IEPs), which are formal written special education plans that consist of goals and objectives to maximize the student's educational achievement (Brady). Chronically ill schoolchildren may qualify for IDEA if they are in need of special education services or it is determined by the school district that they are unable to progress effectively in regular education as a result of their disability (Shapiro & Manz, 2004). Children with chronic medical conditions who receive IEPs typically have severe or multiple needs and are provided accommodations after intensive pre referral interventions and psychoeducational evaluations (Clay, 2004; Shapiro & Manz). Students with illness-related IEPs are often categorized as being Other Health Impaired (OHI) and require both medical and educational interventions due to learning difficulties, such as loss of cognitive functioning after chemotherapy (Shapiro & Manz). The current LQTS guidelines will not outline the details for the IEP, as many students will not have comorbid learning disabilities, but it is important to note that IEPs are designed to create

well-integrated goals that take into account a child's medical and educational needs (Phelps, 2006).

The Rehabilitation Act of 1973, otherwise known as Section 504, is a civil rights law mandating that all schools receiving federal funding meet the educational and medical needs of children with disabilities, including those with chronic illnesses, within the regular education classroom and without identifying them as eligible for special education (Brady, 2004; Shapiro & Manz, 2004). Hence, Section 504 is federally regulated and applies to disabled students who are ineligible for special education services (Brady). Known as 504 Plans, these legally binding formal written prescriptions for interventions ensure that schools do not discriminate against people with disabilities, providing free and appropriate education (Clay, 2004; Phelps, 2006). Unlike the strict IDEA categories of eligibility, under Section 504, a person is considered to have a disability if they have a physical or mental impairment that substantially limits one or more major life activities and has a record of the impairment (Brady). Although determining if a disability substantially limits a major life activity is fundamentally subjective, Smith (2002) recommends that school personnel consider the severity, length, and long-term impact of the condition. If a school finds a student with LQTS to be ineligible for services, parents can either request a copy of the district's 504 procedural safeguards, documents which provide federal 504 policies and grievance procedures, or request a due-process hearing through the school district (Brady). In addition, unlike IDEA, Section 504 does not provide funding to districts for their compliance, although schools are still required to provide related services, affording disabled children with equal opportunities for school achievement (Lee & Janik, 2006). Lastly, under Section

504, there are no formal procedures for referrals or accommodation coordination, leaving each school to independently determine their referral, documentation, and implementation processes (Brady). In general, school principals, nurses, or school counselors are often the primary contact people for school-based accommodations, or know the appropriate person to contact. Unfortunately, the lack of formalized procedures can leave families without much structure or guidance.

General accommodations for youth chronic illness may include academic support after school absences, allowing children short breaks in school, and providing preferential seating in the classroom. For children with LQTS, plans should be individually customized, based on the assessment results and collaborative interviews mentioned previously. Even with children who have the same chronic illness, different services and types of interventions may be necessary (Phelps, 2006). Therefore, the interventions should be designed through multidisciplinary collaboration, taking into account their unique developmental, medical, mental, social, and educational needs. Similar to IEPs, parents have the right to participate in meetings, involve others in the meeting, stay informed of their child's school progress, give or revoke consent for evaluations, request an independent educational evaluation, and disagree with the school's recommendations (Phelps). Although this paper will not detail other forms of accommodations, individualized health care plans and emergency care plans are also strategies to manage school-based health care and emergency needs (*SADS works to keep children safe at school*, 2009). As most adolescents with LQTS will qualify for a 504 Plan, this research will focus on the process of obtaining and creating 504 Plans. The steps in creating an LQTS accommodation plan are outlined in Appendix A.

Step 1: Awareness/referral. The first step in creating an LQTS accommodation plan is to make it known to the school that LQTS services are needed, known as a pre referral (Shapiro & Manz, 2004). Parents, medical professionals, psychologists, students, and even school staff can contact the school to make them aware that a student has LQTS, necessitating an assessment for medical accommodations. Schools can be made aware at any point in the year, but it is optimal to discuss the prospect of an LQTS accommodation plan, or request a yearly review of an existing plan, at transition periods, such as the beginning of each school year, as soon as a child receives the diagnosis, or as soon as a student moves into a new school district. These plans should be updated yearly, as the students' needs may change and the school's personnel and resources may differ year to year. All teachers, including substitutes and those teaching enrichment and supplemental courses, should be given a copy of the 504 Plan.

The referral source should consider what they want to see happen, such as gym class accommodations, their view of the problem, including LQTS characteristics, what type of help they would like to receive, such as nurse availability, who they would like to be involved, including a medical professional, and the desired outcome, such as reduced risk and improved emergency preparedness (Carlson et al., 2004). Once this initial contact has been made, schools usually require a written request from the child's parents or legal guardian for a medical evaluation, details regarding LQTS, including a description of the condition, associated risks, and the ways the condition adversely affects the child's educational performance, documentation of the child's health condition from a qualified medical professional, such as a pediatrician or cardiologist, and permission to conduct an assessment (Phelps, 2006).

Step 2: Evaluation/assessment. Once the school district determines that the child's illness interferes with school, deciding that 504 accommodation services are necessary, the second critical component in developing a school-based LQTS accommodation plan is to conduct a comprehensive biopsychosocial assessment of the student's medical, social, educational, and emotional needs (Power & Parrish, 1995). In order to do this, schools typically have generic forms for parents to sign, permitting the school to conduct the evaluation and communicate with other professionals, known as a release of records. Next, schools can conduct informal assessment interviews or administer formal standardized measures to evaluate the child's psychosocial, medical, and educational functioning. According to Rodrigue, Gonzalez-Peralta, and Langham (2004), examples of biopsychosocial assessment domains may include physical health history, quality of life, development, cognitive functioning, academic achievement, psychological coping, medical adherence, and family functioning. Other sources of evaluation may include teacher observations of peer relationships, previous school medical and educational records, psychological test results, child interviews, parent interviews, teacher interviews, medical professional interviews, medical record reviews, child-self report rating scales, teacher rating scales, and parent rating scales (Clay, 2004). These detailed assessments are often conducted as a result of a 504 request, leading to appropriate and comprehensive accommodation plans. Since adolescents with chronic illnesses may be in need of additional psychological interventions, assessments for anxiety, depression, and coping should be conducted, which can also be performed by contract clinical psychologists, depending on the school's resources and needs. Domain-specific self-report rating scales, including widely utilized screening instruments that assess

behavioral and psychological functioning are often used (Simonian & Tarnowski, 2004). Health-related quality of life instruments are also available, including the Pediatric Quality of Life Inventory (PedsQL) (Varni, Seid, & Rode, 1999). Checklists of general stress can also be administered, including questions pertaining to declines in grades, general mood, worries, anger, and fatigue (Clay). These assessments should be conducted periodically, as determined collaboratively by the intervention team, and should include multimodal and multi-informant sources of data in order to comprehensively address the student's needs.

Since LQTS is a heterogeneous syndrome, it is important for a detailed and specialized assessment interview to be conducted, specifically related to the student's triggers, risks, and type of LQTS. According to Clay (2004), student interviews are a critical source of information when formulating accommodation plans and should include relevant academic, family, medical, psychosocial, behavioral, social, and cultural history. It is important to ask youth about the reasons for the assessment, problems they may be having associated with LQTS, the contexts in which the problems are occurring, coping strategies they use, and possible solutions (Clay). Based on this study's hypothesis and literature review, when interviewing adolescents with LQTS, questions of particular salience include perceptions of social restrictions, co curricular involvement and interest, type of LQTS, triggers, fears, medical events, medications, previous school accommodations, and prior medical and psychological treatments.

Adolescent questions related to self-esteem (SE), based on the literature review, should pertain to feelings of social, athletic, and overall competence. For instance, a relevant question could be if the child believes they are as good in sports as they would

like to be. If they respond that they are not, a follow-up question could ask if they attribute their lack in athletics to their personal skill sets or to their syndrome. When thinking about the social comparison research, adolescents could be asked how they view themselves compared to others their age, such as their ability to make and keep friends, their connectedness to school, their opportunities to engage in school activities, their involvement in developmentally appropriate social activities, the number of friends they have, and their opportunities for the future. Questions assessing SE should address the student's thoughts, feelings, and behaviors related to their feelings of self-worth and self-satisfaction. Formal SE assessment information can also be gleaned by administering batteries such as the Self-Perception Profile for Adolescents (Wichstraum, 1995). In addition to questions related to overall SE, it would be useful to ask students questions pertaining to the impact of LQTS on their school adjustment. For example, a counselor could ask an adolescent how students and staff treat them, followed by asking if their positive or negative interactions are attributed to situational or dispositional characteristics, such as having LQTS. Additionally, inquiring if the adolescent has sufficient opportunities for involvement in extracurricular and social activities could be an indicator of their general school connectedness and satisfaction.

In addition to SE assessments, it is important to ask adolescents about their knowledge and perception of LQTS, in general. For instance, it would be useful to ask if they are knowledgeable about their own condition and their illness perceptions. If a child is unaware that he or she could die from the condition, it will be critical for school staff to describe LQTS consistent with the family's description. Additional questions could ask if the child has witnessed a cardiac event before, such as that of a relative or parent, if they

have been symptomatic in front of their peers before, the impact of that incident, and if they know others who have LQTS and their outcomes. These questions can help assess illness-related fears and feelings of vulnerability. Asking the adolescent when their condition was first diagnosed, if they ever had a symptomatic episode and the circumstances of the event, and the frequency of their medical events can also help to guide the school accommodation plan and provide a greater understanding of the child's unique circumstance and beliefs. Lastly, assessment interviews should also have strength-based components, including the student's supports, positive coping, personal assets, sources of strength, and close family contacts. The ultimate goals of assessments and interviews are to guide the creation of appropriate LQTS accommodation and intervention plans, helping the schools aim to improve things for the child.

Focusing on a collaborative model, parents are also a critical source of information when creating LQTS accommodation plans. Parents can provide their perceptions of the adolescent's functioning, and their illness-related perceptions may differ from that of the adolescent. Parents should be specifically questioned about their concerns regarding their child's health and well-being in school, and differences between parent's views and their child's should be discussed openly. For instance, illness-related questions could be about how often and what type of medications the adolescent takes, if the child wears a medical identifier, if the school needs to administer medications, if the child experiences medication side effects, and if they typically adhere to the regimen. Additional questions to guide interventions can include whether the child has food restrictions, such as caffeine, if they are compliant, if the child needs breaks from physical activities, when resting is important, what accommodations have worked for the

child in the past, if they have a pacemaker or implantable cardioverter defibrillator, if the child has any other known medical or mental health problems, if they have preferred medical contacts, and if they have any other school-related problems, such as academic or social. It is also important to ask parents about their beliefs regarding the LQTS and their family's coping styles, as those factors may influence the amount of school-based support that they will seek (Carlson et al., 2004). Lastly, it is important to inquire about what they feel comfortable disclosing to other students and staff (Clay, 2004). Often, schools will provide parents with a consent form for illness disclosure, detailing what can and cannot be shared with others (Clay).

Maintaining a consultative approach to the creation of intervention plans, it is beneficial for individuals with a range of skills, knowledge, and expertise to collaboratively share information and problem solve (Sheridan & Cowan, 2004). Therefore, it is optimal for parents of adolescents with LQTS to sign a release of records, allowing medical and mental health professionals who are knowledgeable about LQTS to collaborate in the development of the intervention plan. Asking medical professionals for collaboration can help the intervention team to brainstorm emergency plans and identify and prioritize the most critical needs, enhance problem solving, and provide medical expertise (Sheridan & Cowan). Medical professionals, such as a cardiologist or the child's primary care doctor, should be asked any unanswered medical questions, inquiries such as how LQTS affects the child's functioning, what curriculum must be modified, what emergency devices should be purchased, the steps for a crisis plan, and a list of preferred doctors, medical professionals, or hospitals in case of an emergency. The child's doctor can make recommendations pertaining to restrictions in physical activity

and medication management plans, explain the roles of various doctors that care for the child, and help make clear what LQTS is and how it is treated. Licensed psychologists who have expertise in adolescent chronic illness can also advocate for the child's social, psychological, and emotional needs. Lastly, collaborating with professionals can also provide excellent resources for staff training in emergency preparedness and equipment training, such as with AEDs and psychological crisis management. Sample interview and assessment guidelines are outlined in Appendix B.

Step 3: Creating the intervention plan/team conference. First, when creating a 504 Plan, it is most advantageous for a multidisciplinary meeting to be held. Often, schools have a 504 Plan coordinator/liaison who is the primary contact person and determines eligibility for services, sets up multidisciplinary meetings, creates the accommodation plan, and serves as the liaison between the child, school, parents, medical professionals, and related agencies. It is important to request the 504 meeting, in which there is a conversation regarding the degree to which LQTS affects the child's school performance, including information from the child, parent, teachers, physical education specialists, and medical and mental health professionals (Phelps, 2006). Actively collaborating in the development of the plan improves follow-through, accountability, and comprehensiveness. During the 504 Plan meeting, it is critical for the team to review the assessment data and mutually problem solve, determining what accommodations will allow the child to have the same educational opportunities as others and delegating roles for implementing the accommodations. The literature indicates that intervention plans should acknowledge the family's perspective, concerns, level of involvement, desired outcomes, roles and responsibilities, and steps to achieve goals (Carlson et al., 2004).

These multidisciplinary meetings should be held at least once per year, as early as possible, to review the child's progress and needs, while communication between the school, parents, child, and medical professionals should occur on an ongoing basis (Clay, 2004).

Although it is not an individualized or comprehensive listing, the following accommodations may address some of the psychosocial and self-esteem needs of adolescents with LQTS: Providing the child with alternate gym class activities (as determined by medical professionals), making revisions to extracurricular activity requirements (as approved by medical professionals), permitting the child to change for gym in an alternate location if there is scarring from surgery and it is his/her preference, allowing the child to leave class at determined times to take medications with the school nurse, permitting the child to take breaks from physical activity or rest if medications cause fatigue, allowing the child to cease physical activity if they deem it necessary, providing supervision at all times during physical activity, creating a buddy/peer partnering system if requested, monitoring fluid intake to maintain hydration in hot climates and temperatures, maintaining adequate food intake to sustain energy and medicine, such as allowing the child to eat snacks, monitoring restrictions to foods or substances, such as those containing caffeine or chocolate that increase sympathetic arousal, providing staff trainings, developing an emergency plan, purchasing medical devices such as AEDs, ensuring existing AEDs are in an accessible location and operating properly, displaying signs throughout the building indicating where the AED is located, providing support groups and psychological counseling as needed, communicating health-related concerns or breaches of the 504 Plan with parents, such as

if the child was noncompliant with their gym restrictions or missed medications, maintaining ongoing communication with parents and medical specialists, monitoring vital signs as needed, observing student responsiveness as needed, and arranging for nurses to be available during school, after school, and on school trips. For field trips, an AED, an individual trained in CPR, and AED use, and first aid should be available, along with the student's emergency contact information, an emergency plan, and a communication device, such as a cell phone (*SADS works to keep children safe at school*, 2009). For encouragement with medical adherence, the 504 Plan can include such strategies as monitoring barriers to treatment adherence on a monthly basis, such as discussing with the adolescent their difficulties and successes in abiding by gym class restrictions (Phelps, 2006). For adolescents with LQTS, a useful approach to maintain their self-esteem could be offering alternate gym activities that allow them to stay involved in the gym class, such as creating noncompetitive recreational activities (as approved by their doctor), allowing the student to keep score of games, offering the child the opportunity to referee or coach, or even permitting them to take roll for class. If an adolescent with LQTS is required to take medication at school, the school can assist the student with creating a chart that tracks their adherence, or writing prompts, such as daily reminders, in their school calendar. As disruption is associated with medication compliance, adolescents with LQTS may prefer taking their medications during breaks, as opposed to leaving during class (Power et al., 2003). Within each of these strategies, collaborative team members should develop ways to maximize school involvement and minimize the adolescent's feelings of dissimilarity from others.

The accommodation plan may also include the development of a medical management plan. These plans usually require parents to fill out a student health information form, provided by the school nurse, which is given to all staff. Included on the form is the student's name, medical condition, triggers, medications taken, emergency contacts, physician's name and contact information, where emergency equipment is located, such as the AED, and steps in the event of an emergency, such as determining their heart rate, calling 911, and contacting the student's parents if a child has a fainting spell or cardiac arrest. Another 504 Plan prevention strategy could be contacting local hospitals, police stations, emergency medical technicians (EMTs), and emergency health facilities to notify them of the student's condition in the event of a crisis. Detailed in the plan should also be who is responsible for implementing the 504 strategies and how often they are to be employed. Lastly, after the meeting, the final 504 Plan should be written, signed by all parties present in the meeting, and distributed to all school staff responsible for the child's well-being, including teachers, administrators, counselors, aids, bus drivers, and lunch aids. The 504 Plan should be kept on file, usually with the school's administration or guidance office, for documentation and future reference. A sample LQTS 504 Plan is provided in Appendix C.

As an important component of any medical intervention plan, the literature indicates that it is critical to discuss with the child and his or her family their comfort with disclosing information about their illness (Clay, 2004). Some adolescents may be hesitant to share information with teachers and peers about LQTS, fearing embarrassment. Therefore, it is necessary for the intervention team to discuss with the child, the family, and medical providers what they would like to disclose versus what

they need to disclose to others (Clay). After collaboratively determining what information can be disclosed, it should be clarified when to disclose information, who can know what, and who should be involved in the disclosure process (Clay). For instance, an adolescent with LQTS may only want his or her immediate teachers to know about their condition. Although it is important to consider the adolescent's comfort with self-disclosure, it has been recommended that all staff involved with the child's activities be made aware about their condition, in order to provide consistent services and maximize safety and welfare (Rae & Fournier, 2004). Ultimately, the benefits of disclosing information to everyone in the school should be weighed against the risks and should be mutually agreed upon by the student, parents, school, and medical consultants. Lastly, confidentiality and the limits to the student's confidentiality (if a danger to themselves or others) should be discussed in the accommodation plan meeting (Rae & Fournier, 2004).

Clay (2004) also suggested that school personnel talk to the adolescent and the family about questions typically asked by peers and staff, such as if LQTS is contagious, if the teen will die from it, and why the adolescent has different sports activities. Then, the adolescent and the parents should formulate answers that they are comfortable with, preparing for replies when asked questions. Optimally, the child should write down the answers to commonly asked questions and return them to school personnel, in order to document what they feel comfortable disclosing. If the adolescent decides to change his or her boundaries of confidentiality and inform additional staff, the accommodation plan should be changed accordingly (Rae & Fournier, 2004).

Reentry programs are also an important part of an intervention plan for adolescents with chronic illness (Anderson, Taylor, & Boevig, 2004). These plans assist

with educational, emotional, and social transitions back to school, attempting to maintain school connectedness and academic progress. As a part of the 504 Plan, a school reentry program should be established, preparing for smooth transitions back to school after an illness-related absence (Clay, 2004). Although schoolchildren with LQTS may not be absent for extended periods of time, reentry plans are often created as a precautionary measure.

When creating reentry plans, Clay (2004) recommends that parents inform the principal of the child's absence and when the child is returning to school, contact the school nurse, and talk to the school counselor to discuss any needed psychological services. If a student undergoes surgery, it is important to discuss discharge instructions, including any additional athletic restrictions (*SADS works to keep children safe at school*, 2009). For an adolescent with LQTS, it may be useful to select a faculty liaison who can speak with the student's teachers and classmates, educating them about the syndrome during the child's absence, answering their questions and concerns, and informing them about potential restrictions once the child returns. Common questions may be if the child will require another absence and what they should say upon their return (Anderson et al., 2004). It is also useful to designate a school-based point person to maintain contact with the adolescent while they are absent, sustaining school connectedness. Since school connectedness has been associated with lower levels of psychological problems and violent behavior, and personal satisfaction is often derived from school-based accomplishments, it is important for students to sustain school connections during extended absences, maintaining their feelings of self-worth and achievement (DuBois et al., 1998; Hall-Lande et al., 2007). Additional accommodations include arranging for

homebound instruction if needed, providing an extra set of textbooks, modifying the curriculum, and informing parents about the child's progress once they return to school.

Step 4: Psychological interventions. Since adolescent chronic illness has been associated with difficulties in psychosocial functioning and self-esteem, it is important for schools to provide appropriate empirically supported cognitive-behavioral psychological services to those in need (Shapiro & Manz, 2004). School-based mental health services are beneficial, as they are accessible, relationships can be easily formed, and they are economical. If a licensed clinical psychologist is collaborating in the development of the accommodation plan, he or she may be able to provide guidance in implementing psychological services, such as training staff on empirical techniques and ensuring that the treatment is delivered in the most ethical and efficacious manner. Within the school, counseling sessions can be provided by a school counselor or school psychologist and may include topics deemed necessary from the assessment and interviews, such as self-esteem, peer relationships, bullying, living with a chronic condition, illness uncertainty, coping, stress management, and school integration (Clay, 2004; Phelps, 2006). For instance, in his book, Clay provides tips and intervention worksheets to help youth cope with the impact of a variety of chronic illnesses. Based on the student's needs, counseling sessions can also focus on health promotion, healthy development, and treatment adherence. The frequency of the sessions can be determined during the 504 Plan meeting and can range from periodic check-ins to frequent meetings. If the child experiences an event during the school year, it should be recommended that he or she be referred to the school counselor or psychologist for counseling and an assessment, determining if the student is in need of additional services. In order to ensure

that the treatment is tailored to the needs of the adolescent, it is most beneficial to base interventions on the previous assessment and interview results, either in an individual or group format. For instance, Phelps suggested that providing opportunities for youth with chronic illnesses to get together with others with similar experiences can promote self-esteem, and support-groups can promote treatment adherence. Group interventions can also help to reduce stigma and increase comfort in disclosing illness-related information (Clay). Alternately, schools should first assess what group the adolescent identifies, as he or she may not consider himself or herself disabled or impaired. Therefore, putting the adolescent with other students who have visible disabilities or other health problems may be upsetting. Lastly, school counseling services can connect adolescents with LQTS to outside agencies and support groups, such as LQTS online user groups and cardiac organizations.

Step 5: School-based trainings. School-based trainings are a critical component of an accommodation program for adolescents with LQTS, focusing on primary and secondary prevention efforts. As staff may be called upon to conduct life-saving interventions, it is vital that they are well educated about LQTS. Power et al. (2003) suggested that teacher education programs, including faculty workshops and presentations, include instructions about the medical condition, describe challenges associated with the syndrome, and present strategies for dealing with them. Trainings should also include a basic understanding of the syndrome, how to identify a medical emergency, and whom to contact for questions or in the event of an emergency (*Diabetes at school*, 2009). Therefore, for LQTS, it is necessary to explain the 504 Plan, triggers and symptom displays, as described in the introduction portion of this manuscript and

emergency management of the syndrome, such as calling for help and dialing 911 if fainting occurs (*SADS works to keep children safe at school*, 2009). As practical suggestions, DeMase (2004) recommended that school staff be reminded that cardiac illness does not define the child, and parents should be reminded that school staff may have fears associated with treating a child with a chronic condition. Power et al. (2003) stated that peer education programs, aiming to improve peer support, may be useful, but should be weighed against the potential for social isolation and self-consciousness.

Maintaining the collaborative model, a specialist in LQTS, such as a cardiologist or physician, the school nurse, parents, and the adolescent should be involved in the creation and execution of school trainings, and should ideally include all staff and faculty who interact with the student. Although not often possible due to shortages in school resources and staff availability, it is optimal for all school employees, including teachers, administrators, aides, lunch staff, substitute teachers, and even bus drivers to be educated about LQTS, emergency plans, treatment regimens, the use of AEDs, cardiopulmonary resuscitation (CPR) and first aid, implanted cardioverter defibrillators (ICDs), symptom displays, and possible medication side effects. It has been recommended that at least two employees per building, and ideally two employees per floor, have current CPR and first aid training through a nationally recognized training organization (*SADS works to keep children safe at school*, 2009). It is also important to talk to the staff about the child's accommodation plan, explain the rationale for restrictions, and provide them with LQTS resources and references. It is ideal for these training programs to be conducted at the beginning of each school year to teach new staff and retrain existing staff. It is also important for an individual involved in the training to be available for faculty questions

or follow-up trainings throughout the year. It has even been recommended that all employees receive a picture of the child for safety and identification purposes (*SADS works to keep children safe at school*, 2009). Given the delicate developmental period of adolescence, this intervention should be approved by the student and their family.

Step 6: Emergency response plans. Although the LQTS 504 Plan addresses primary, secondary, and tertiary prevention and intervention models, it is necessary for schools to implement emergency response procedures to maximize student health and safety, improve communication with emergency responders and parents, and initiate post trauma information and support. Crisis response teams are typically comprised of the school's principal or vice principal, school counselors, school psychologist, school nurse, and teachers trained in crisis management. These general roles and emergency steps are usually delineated, in full detail, within the district's emergency manual. The complete details of emergency response plans will not be covered in this thesis, as they should be specific to each case and situation. Alternately, a suggested sequence of steps specifically for adolescents with LQTS is provided.

First, it is important to establish a crisis team, if one has not been already established, including school staff who volunteer to care for the adolescent with LQTS in the event of an emergency. Second, these individuals should receive additional specialized training, conducted by a medical specialist, such as what to do if the student is unconscious, how to contact the school nurse, when to call 911, how to contact the student's physician and parents, steps to reach emergency contacts, what to do if the student's ICD administers a shock, how to use the AED, when to use the AED, and what to do until the ambulance arrives (*SADS works to keep children safe at school*, 2009).

This plan must be written, signed by the 504 Plan committee, and distributed to all staff. For instance, if a student with LQTS fainted, a sample emergency sequence would be calling 911, notifying the school nurse and principal, administering CPR if needed, retrieving and using the AED if needed, and calling the student's parents (*SADS works to keep children safe at school*, 2009).

Next, once the emergency sequences have been established, members of the crisis team should be assigned specific roles, such as coordinating and alerting the crisis response team, removing other students and nonessential people from the situation, calling the student's parents and medical doctor, calling for an ambulance, administering CPR, getting the AED, using the AED, directing emergency personnel once they are in the building, briefing emergency responders, handling the media, and assisting the nurse. These roles should be assigned to staff for the regular school day, after school, and while on school trips. As mentioned previously, these emergency response plans must be tailored to the student's unique circumstances and established during the 504 Plan meeting. Ideally, each crisis response member should be present at the meeting.

Step 7: Disaster preparedness plans. In addition to emergency response plans, schools have district wide disaster preparedness plans that detail how to notify staff, students, and parents of an emergency or the death of a student. Since these plans are typically detailed and specific to each school district and incident, this manuscript will provide an outline of the procedures applicable to a student with LQTS. As with the emergency response plans, designees should be appointed, including individuals to secure facts, notify the crisis response team, inform students and staff, provide support, obtain

funeral and memorial information if applicable, and manage the media. A flowchart of these procedures is provided in Appendix D.

First, as soon as possible, all facts need to be secured, including what happened, whom it happened to, and the outcome. Often, the school principal is designated to verify information, notify the superintendent, contact the school's crisis management team, develop letters to the faculty, students, and parents, and initiate the emergency phone chain. Remaining mindful of confidentiality and respect for the family, the child's parents are often asked what they would like disclosed to the faculty, students, and other parents and about details regarding the funeral and memorial, if applicable. At this time, principals often determine if substitute teachers are needed to relieve classroom teachers.

Second, the faculty should be informed about the emergency or tragedy, which is optimally before students receive information. If the crisis occurs after school hours, staff are frequently notified via a phone chain initiated the night before by the principal. These phone calls notify employees of the emergency and about a mandatory meeting the next morning. During this emergency meeting, employees are provided with the facts of the situation, plans for disclosure, changes in the daily schedule, and possibly funeral information, and staff support is offered. Alternately, if the emergency or loss occurs during the school day, crisis response designees usually contact the student's parents and speak personally with teachers throughout the day, distributing memos with the above factual information. In these circumstances, meetings are often held at the end of the day to provide faculty with additional information and assistance. Support staff, such as secretaries, should be given scripts to read if asked questions or receiving phone calls from students or parents. If relatives of the student live in the school district, the faculty

at that school are usually contacted as soon as possible, and similar disaster response procedures are implemented. Often, it is prearranged that volunteer counselors and crisis teams at neighboring school districts will assist in the event of an emergency. A sample announcement to staff is provided in Appendix E, including the details about the event and funeral arrangements if applicable.

Next, students should be informed of the incident and rumors should be dispelled, usually during class by a teacher. Again, this is often conducted by a designee, and consistent information is provided to all students at the same time. A sample announcement to students is provided in Appendix F, including the facts of the situation, normalization of reactions, how to respond to the media, and opportunities for counseling. Students are typically encouraged to talk about the situation with their teachers, and group and individual counseling is usually offered by the school counselor, school psychologist, and crisis team members. Classmates who were particularly close with the student, individuals who witnessed the incident, or other vulnerable students should be observed and offered additional psychological services.

Parents should also receive information about the crisis, including the facts of the situation, available student psychological services, how to address the media, and funeral information if needed. This information is typically disseminated in a memo created by the principal (Appendix G). Often, these pamphlets are mailed or given to students to bring home. Then, students, parents, and faculty should be provided with resources and information pertaining to reactions to crisis or grief, including when psychological services are needed and how to obtain them. At the end of the day on which the crisis is

first disclosed, the principal, guidance team, and the crisis response team typically meet to determine if the school community needs additional crisis services.

As an important component of a disaster preparedness plan, debriefing normally occurs a few days after the incident, during which the situation is processed with students and faculty, and funeral and memorial information is provided if needed. Lastly, throughout the next few months after the incident, teachers are often asked to refer high-risk students, such as those who witnessed the event, had relationships with the student, or have preexisting mental health conditions, to the crisis team, and continued student and staff discussions are encouraged. Consultation with a clinical psychologist can assist in training staff about the warning signs for anxiety, depression, and grief and provide education regarding effective psychological treatments. Empirically supported treatments for adolescent anxiety include exposure tasks, behavioral modification, systematic desensitization, behavioral rewards, anxiety hierarchies, and modeling (Kazdin & Weisz, 1998). For instance, a well-established cognitive-behavioral therapy (CBT) for anxious children is the Coping Cat program, a 16-session program which includes role plays, relaxation training, exposures, cognitive restructuring, and reinforcement (Kendall & Hedtke, 2006). Supported components of adolescent depression include psychoeducation, emotional regulation, enhanced social interactions, social skills training, relaxation training, cognitive restructuring, and problem-solving training (Kendall & Hedtke). According to Armstrong and Briery (2004), adolescents who lose a friend generally report wanting support from parents and teachers for 1 to 3 months. They should be given adequate time and opportunities to process the bereavement, memorial and remembrance activities can be helpful, and psychological professionals should be included. Therefore,

well-established disaster preparedness plans are critical for student and faculty well-being, supporting those impacted by the event and preventing the onset of additional crises. Although these plans are typically detailed in each school district's faculty manuals, they may not be reviewed frequently by employees and parents may not be aware of them. Hence, it is important to collaboratively discuss these procedures during the 504 Plan meeting, comprehensively addressing the potential needs of the student and this or her family.

Implications

The implications for providing guidance in the creation of comprehensive LQTS accommodation plans are far reaching, and may lead to greater quality of life, minimize the impact of LQTS at school, improve self-esteem and psychosocial functioning, increase LQTS awareness, and assist parents, schools, and health professionals in advocating for the needs of adolescents with LQTS. Often, schools and families are unsure about how to initiate school plans, what the child qualifies for, what are appropriate recommendations, and who is responsible for the accommodations. It is intended that the procedures provided in this manuscript will guide families and school districts, from start to finish, in developing safe and effective LQTS accommodation plans. The recommendations for these plans are potentially lifesaving, making them a critical part of a child's educational plan. If the suggestions provided can make a difference in the life of only one child, this manuscript has accomplished the goal it set out to achieve. Since the suggestions provided are the optimal recommendations based on the literature, many children will not need all of the services recommended, nor will all

schools be able to implement all of the options. Therefore, collaboration and mutual agreement are critical when individualizing the plan to the needs of the adolescent.

Since LQTS is a relatively rare condition, another aim of this thesis is to educate professionals who work with the LQTS community, leading to increased awareness, research in the areas of needs assessment and advocacy, and future studies. Even though the data is not presented as proposed, simply increasing awareness about the condition and educating others about the strengths and needs of adolescents with LQTS is progress within the community. Moving forward with the creation of LQTS prevention and intervention strategies will hopefully sensitize practitioners to the implications of living with LQTS, providing the population with greater support, advocacy, and health.

CHAPTER 6

Discussion

Findings

Based on the current literature indicating that adolescents' self-esteem (SE) is often impacted by the effects of chronic illness (CI), and long QT syndrome (LQTS) is a chronic and potentially life-threatening genetic cardiac illness, this thesis hypothesized that the unique challenges that LQTS imposes on adolescents impact their social functioning, school performance, and SE. Specifically, it was hypothesized that adolescents with higher ratings of self-reported school and social functioning would have higher levels of SE. Additionally, it was hypothesized that social and co curricular restrictions, school accommodations, age of diagnosis, frequency of medical events, social functioning, and school performance would predict ratings of SE.

Although the patient sample was not secured within the allotted time for the proposed study, literature pertaining to adolescent chronic illness and LQTS studies supports the need for school-based accommodation plans and crisis response procedures. This is among the first manuscripts to address the psychosocial functioning and self-esteem of adolescents with LQTS. Recommendations provided in this thesis may guide medical and mental health professionals in identifying the strengths and weaknesses in the psychosocial adjustment of youth with LQTS. These strategies will hopefully assist helping professionals to begin to identify some of the psychological and social needs of youth with LQTS and lead to the creation of individualized accommodation plans.

*Future Directions**Feasibility and Acceptability*

As this is one of the first papers to provide comprehensive recommendations for the creation and implementation of adolescent LQTS school-based accommodation plans, it would be useful to conduct research pertaining to the feasibility and acceptability of the model. For instance, a random sample of principals and counselors could receive the outlined plan, and researchers could request they fill out a questionnaire asking if the accommodations are viable and sustainable, given their school's resources and abilities. Parents and adolescents with LQTS could be asked to answer questions related to the suitability of the plan, given the child's medical, mental health, and school, emotional, psychological, and developmental needs. Medical and mental health professionals who work with the LQTS population could also be asked to answer questions pertaining to the comprehensiveness and appropriateness of the interventions. Once feedback has been received and analyzed, the model can be revised and adapted.

Details

Although the 504 Plan recommendations provided are an overview of the steps to create and implement school-based services, there are further details that future studies can provide. For instance, researchers can create additional intervention materials, such as more detailed structured assessment interviews specific to adolescents with LQTS, their parents, and medical specialists. It could be helpful to outline the specific assessment batteries that schools should administer, including cardiac-specific modules and measures specific to chronically ill adolescents. Although this thesis discussed the importance of emergency response plans, future research should detail emergency steps

for a variety of circumstances, such as if a child does not have an ICD, if they do have an ICD, if they are unconscious with a pulse, or if they are found without a pulse. These plans should be created in collaboration with an LQTS specialist to maximize health and safety. Future studies can also describe alternate accommodation plans in more detail, including IEPs and health plans, as adolescents with LQTS may qualify for those services. It could also be beneficial for future publications to create step-by-step LQTS training programs for schools to utilize. As many schools will not have access to a medical professional specializing in LQTS or have the resources to hire a consultant yearly, purchasing the training curriculum for school wide trainings could be cost effective, feasible, and sustainable.

Troubleshooting

Future studies can also provide troubleshooting suggestions, such as if the school does not have a full-time nurse, if the 504 Plan meeting is not collaborative, if school staff is resistant, if there is lack of follow-through with the plan, or if the school does not think the child qualifies for services. Another setback can be if the child's medical doctors are unable or unwilling to collaborate on the 504 Plan. If parents do not want LQTS services, placing their child at risk, if the child is resistant to the accommodation plan, if the school is unable to provide needed medical care, or if LQTS medical specialists are not local to the school are potential challenges that could be addressed in future studies. If medical specialists are unavailable, one suggestion could be to create the plan with the school and then take it to the child's doctor for approval or recommendations or create the plan with involvement from the school nurse (*SADS*

works to keep children safe at school, 2009). If the child is resistant, another suggestion would be to involve only those faculty who are in direct contact with the child on a daily basis. If parents are concerned about school follow-through, they can request follow-up trainings and periodic check-ins with responsible parties to improve their accountability and buy-in. Lastly, future studies should investigate the utility of various empirically supported psychological interventions for adolescents with LQTS, including intervention content, topics, or even manualized treatments. Overall, it was the intention of this manuscript to raise general awareness regarding the steps to secure and implement school-based LQTS accommodations. Future studies should continue to investigate the utility of the recommendations, providing additional materials and resources.

Advocacy

In addition to the refinement and expansion of school-based accommodation plans, there is also a great need for advocacy for individuals with LQTS and their families. In particular, psychologists are well positioned to advocate for individuals in need of psychological and medical services. Advocacy includes helping those who are underserved, such as those with chronic illness, to access care, promoting equality and social justice, and supporting public policy initiatives (Lating, Barnett, & Horowitz, 2009). For instance, the National Council of Schools and Programs of Professional Psychology, whose mission is to promote the improvement and enhancement of the field of professional psychology and its educational programs, includes advocacy as a core competency, highlighting the importance of understanding sociocultural diversity, using liaisons to support clients' needs, influencing psychology policies and regulations,

increasing funding for psychology education, and educating consumers about the services that psychology has to offer (Lating et al., 2009). In thinking about ways to advocate for adolescents with LQTS and their families, psychologists can help guide families through the steps of obtaining and monitoring school-based services, consult with schools regarding the best strategies to maximize school success while minimizing illness visibility, and provide families with psychological services, including assertiveness training to empower them to become self-advocates.

It is important to recognize the steps in guiding clinical psychologists to become advocates. First, it is critical to inform clinical psychologists about services that youth with LQTS may be eligible for, including 504 Plans, Individualized Education Plans (IEPs), and health plans. Providing psychologists with this knowledge gives them a context from which to set goals with the family and offer support and guidance. Since navigating through educational systems can be overwhelming to families and met with resistance at times, psychologists can offer encouragement, support, and active collaboration in the process, contacting key school personnel, advocating for the need for services, and modeling persistence and perseverance in obtaining needed services. Once given knowledge regarding the eligibility for services, psychologists can also provide the adolescent and his or her family with psychoeducation regarding the utility of school-based services, the potential impact of LQTS on self-esteem and psychosocial functioning, how accommodation plans can help support psychological well-being, and the characteristics of effective accommodation plans, including a biopsychosocial, multidisciplinary, and consultative emphasis. Lastly, as local clinical scientists, psychologists can help families to monitor the effectiveness of the school-based services,

including tracking and documenting psychological functioning over time and providing schools with ongoing feedback regarding the utility of the accommodations.

Second, as multidisciplinary consultation is a key component of school accommodation plans, psychologists can be invaluable in guiding schools to implement developmentally appropriate, empirically based, and theory-driven accommodations. For instance, having knowledge about social comparison research and theory, clinical psychologists can help to develop school-based strategies that maximize school involvement while minimizing illness visibility. Clinical psychologists can consult with the school counselor and school psychologist, devising group and individual therapy goals, if needed, and providing them with education regarding the psychological impact of living with a chronic health condition. Lastly, psychologists can collaborate with the child's primary care doctor and cardiac specialists, imparting knowledge about accommodation plans, patient eligibility, and psychological impact of the condition, and the cognitive and behavioral components of the plans. This multidisciplinary approach to care provides the families with the most comprehensive and well-rounded biopsychosocial emphasis to treatment. Linking psychologists and school consultants together will ultimately strengthen the impact of advocacy initiatives for adolescents with LQTS and their families.

In addition to actively advocating for the needs of adolescents with LQTS, psychologists can also provide services within therapy sessions to help parents and their children become self-advocates. Self-advocacy includes components such as knowledge of self, communication skills, knowledge of rights, and an understanding of leadership, all of which can become goals within therapy sessions (Test, Fowler, Wood, Brewer, &

Eddy, 2005). For instance, psychologists can provide clients with psychoeducation regarding chronic illness, patient rights, and communication skills training. In addition, psychologists can provide families with assertiveness training to empower them to advocate for effective school-based services, the need for medical consultation and collaboration, and the necessity of monitoring and implementing emergency and health plans. Assertiveness training can also help families to overcome some of the roadblocks they may encounter with school districts, such as lack of follow-through, resistance, and poor implementation. Therefore, clinical psychologists can be an invaluable asset in helping youth with LQTS receive accommodation plans, playing a key advocacy role and providing families with the tools to become self-advocates.

Future Research Considerations

Diversity

Future studies should recruit diverse samples of adolescents with LQTS from a variety of backgrounds and ethnicities, in order to increase generalizability. Research is also needed for the children, adults, and parents of LQTS patients. Given that this is one of the first psychosocial LQTS guideline manuscripts, all other populations and questions of inquiry will add to the understanding of this population. Lastly, future research should longitudinally examine the psychosocial impact of living with LQTS. By tracking patients over time, timelines and causal relationships could inform preventative and treatment strategies.

Moderating Variables

Future studies should employ large samples of adolescents with LQTS in order to examine additional mediating and moderating patient variables that could impact psychological and social well-being. For instance, individual variability, including socioeconomic variables, culture, ethnicity, and gender, exists among LQTS patients, potentially impacting their self-perceptions and social and school functioning in systematic ways (Williams, Holmbeck, & Greenley, 2002). In addition, since research indicates that male and female global SE ratings often differ among healthy populations (Quatman & Watson, 2001), it would be informative to learn if this applies to adolescents with LQTS. Researchers should also analyze and report data on the effects of therapist characteristics, as they may differentially impact subjects' responses to question items.

Multiple Domains

Researchers should investigate the impact of living with LQTS across multiple domains. Since this study primarily focused on the impact of social functioning, school performance, and SE of adolescents, there may be other psychosocial domains that are impacted by the condition. For instance, anxiety levels, depression ratings, and quality of life are three areas that may be affected by living with LQTS. Since adolescence is a time for sexual exploration (Santrock, 2001), and sexual activities are often associated with heart rate elevations, future studies should investigate the impact of LQTS on sexual development and perceptions of risk associated with physical intimacy. Also, future studies should examine overall school belonging in adolescents with LQTS, in order to increase positive outcomes such as academic attainment and school involvement in later years (Pittman & Richmond, 2007). In addition, since schools are an integral component

in the development of adolescent SE, where peer relationships develop, feelings of competence emerge, and academic skills are assessed, research should investigate the involvement of schools in treatment programs, such as the adoption of health behaviors and medication management (Williams et al., 2002).

As indicated earlier, research on painful or physically restrictive chronic illnesses may not generalize to LQTS patients, as they often feel healthy. Therefore, studies investigating the impact of living with diabetes, asthma, cystic fibrosis, and allergies should be replicated using the LQTS population. Specifically, research investigating the impact of athletic and social restrictions on adolescents with asymptomatic CIs would be useful as few studies have investigated these aspects in those with LQTS. In order for medical and mental health professionals to comprehensively understand the needs and strengths of adolescents with LQTS, it is imperative for future research to broaden the scope of inquiries associated with the LQTS population and their families.

LQTS Assessments

Currently, there are few empirically validated LQTS scales that measure psychosocial functioning in patients of any age group. Hence, additional measures are needed to assess the SE, social, and school domains, along with internalizing and externalizing disorders, such as anxiety, depression, and behavioral disorders among LQTS patients. Since the use of existing measures is often preferable to creating new assessments (Kazdin, 2003), future research should investigate the reliability and validity of using current instruments with LQTS patients. For instance, although the Pediatric Quality of Life Inventory (PedsQL) was created and normed using cardiac patients, it was

not specifically developed for the unique needs of LQTS patients. Psychometric properties of the PedsQL should therefore be evaluated in order to ensure that it parallels the customary use of the measure (Kazdin). Lastly, it would be informative to conduct an investigation of the reliability and validity of the personal history questionnaire, developed by the current research team, in order to reduce threats to internal validity posed by instrumentation.

Treatments

Once the literature has established the needs and characteristics of the LQTS population, researchers should investigate the utility of prevention and treatment programs. Treatment plans should be developmentally appropriate, taking into consideration the unique cognitive, social, emotional, physical, psychological, contextual, and behavioral development of LQTS patients (Christian & D'Auria, 1997). Since LQTS is a chronic medical syndrome, it would be interesting to compare treatments that incorporate medical and mental health professionals working as a team, providing comprehensive services. For instance, Christian and D'Auria stated that intervention services that link medical, educational, familial, and peer contexts maximize treatment benefits and sustainability. Adolescent treatments should focus on how to disclose a diagnosis, forming relationships, dealing with difficult peer situations, and handling the negative reactions of others, including school staff and peers (Christian & D'Auria). Prevention programs that include the development of protective factors while considering multiple physical, social, environmental, and psychological risk factors should also be researched (Williams et al., 2002). In addition, psychological screenings could be

implemented as a part of routine medical examinations, including ratings of self-perceptions and SE.

Lastly, future studies should investigate the efficacy of cognitive-behavioral therapy (CBT) with LQTS patients. Optimistic self-beliefs about one's CI often increase health-related behaviors and perceived illness control, and treatment satisfaction is enhanced by good illness understanding (Luszczynska, Gibbons, Piko, & Tekozel, 2004; Veldtman, Matley, Kendall, Quirk, Gibbs et al., 2000). Additionally, individuals who hold general efficacy beliefs about their ability to overcome health obstacles are able to see the relationships among their health behaviors and future outcomes (Luszczynska et al.). Therefore, CBT for LQTS patients could aid individuals in altering misattributions, shaping schemas, objectively evaluating social comparisons, and increasing medical adherence, all of which are associated with psychological, social, and behavioral well-being. Consequently, there may be great utility in CBT as a means of guiding patients to shape their illness outlook and understanding.

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Appendix A

Sample Yearly LQTS Accommodation Checklist

Note: It is best to discuss creating a LQTS accommodation plan, or request a yearly review of an existing plan, at transition periods, such as the beginning of each school year, as soon as a child is diagnosed, or as soon as a student moves into a new school district. These plans should be updated yearly as the students' needs may change and the school's personnel and resources may differ year to year. All teachers, including substitutes and those teaching enrichment and supplemental courses should be given a copy of the 504 Plan.

This checklist lists services that may be needed for a child with LQTS. This is not a comprehensive list of services nor will all services be necessary for each child. Each plan must be individually tailored to the unique needs of each student, and multidisciplinary collaboration is encouraged. This list can be used by parents, schools, or medical providers.

Note: The referral source refers to the person who has made the school aware that a child with LQTS may need an accommodation plan. For instance, parents, medical professionals, psychologists, students, and other school staff can contact the school to make it known that a child with LQTS may be in need of services.

1. AWARENESS/ REFERRAL

- ☐ Make it known to the school that accommodation services are needed (For instance, parents, medical professionals, psychologists, students, and other school staff can contact the school to make it known that a child with LQTS may be in need of services)
- ☐ OR, make it known to the school that a yearly review of LQTS services are needed (Can remove or add previous accommodations)
- ☐ Referral source considers what they want, their view of the problem, type of help that is needed, who they would like involved, and the desired outcome
- ☐ Parents make a written request for evaluation, provide LQTS description, risks, and how it adversely affects educational performance
- ☐ Parents provide documentation of LQTS from a qualified medical professional (Ex: medical script)

2. EVALUATION/ ASSESSMENT

- ☐ Parents sign a permission form for the school to conduct an assessment and evaluation
- ☐ Parents sign a release of records form so the school can communicate with other professionals, such as the primary care physician
- ☐ Parents fill out a consent form for illness disclosure (This details what can/cannot be shared)

- ☐ Parents fill out a student health information form (Provided by the school nurse)
- ☐ School conducts a biopsychosocial assessment of the student's medical, social, educational, and emotional needs. Schools can use clinical interviews and validated standardized measures. Examples of commonly used self-report measures are the Childhood Depression Inventory (CDI) for depression, the Multidimensional Anxiety Scale for Children (MASC) for anxiety, the Youth Self-Report (YSR) for internalizing and externalizing behaviors, and the Self-Perception Profile for Adolescents (SPPA) for self-perceptions (Simonian & Tarnowski, 2004)
- ☐ School conducts a LQTS-specific child assessment interview (Question examples are triggers, risks, social restrictions, co curricular involvement, fears, medical events, medications, previous school accommodations, and medical and psychological treatments. Overall, medical questions related to how LQTS may impact the child's needs at school should also be asked)
- ☐ School conducts a LQTS-specific parent(s) assessment interview (Pertaining to medical needs and health/well-being)
- ☐ School conducts a LQTS-specific medical-specialist assessment interview (Questions related to unanswered medical questions, how LQTS affects the child's school functioning, needed curriculum modifications, emergency devices, crisis plans, activity restrictions, medication management, and a list of preferred doctors, medical professionals, or hospitals in case of an emergency)

3. CREATING THE INTERVENTION PLAN/ TEAM CONFERENCE

- ☐ Parents and/or school requests a multidisciplinary 504 Plan meeting
- ☐ In the meeting, the team discusses how LQTS affects the child's school performance (Optimal to include child, parent, teachers, gym teachers, medical, and mental health professionals)
- ☐ OR in the meeting, discuss progress or setbacks when reviewing how LQTS affects child's school performance
- ☐ In the meeting, review assessment data, problem-solve, and determine accommodations (Ex: ensure the school has operable an AED in an accessible location)
- ☐ In the meeting, delegate roles for implementing accommodations
- ☐ In the meeting, discuss to whom the family gives permission to disclose information about LQTS
- ☐ In meeting, discuss with child, family, and doctor what **must** be disclosed
- ☐ In the meeting, determine what info will be disclosed, when, and who can know what
- ☐ In the meeting, discuss confidentiality and the limits (For instance, breached if a danger to themselves or others)
- ☐ In the meeting, families are made aware of questions typically asked by peers and staff (Prepares student and parents with replies when asked)

- ☐ In the meeting, discuss the prospect of and/or create/modify reentry program to assist with educational, emotional, and social transitions after an illness-related absence
- ☐ In the meeting, discuss the emergency response plan and delegate roles
- ☐ 504 Plan created or modified
- ☐ 504 Plan written and signed by all parties present in meeting
- ☐ 504 Plan distributed to all school staff responsible for child's well-being
- ☐ 504 Plan kept on file with administration or guidance

4. PSYCHOLOGICAL INTERVENTIONS

- ☐ School provides psychological services based on the assessment and interview results (For instance, could be related to self-esteem, peer relationships, bullying, living with a chronic condition, illness uncertainty, problem-solving, coping, stress management, and successful school integration)
- ☐ Clay (2004) offers worksheets and treatment recommendations for schoolchildren with chronic illnesses
- ☐ A well-established cognitive-behavioral therapy (CBT) for anxious children is the Coping Cat program, a 16-session program which includes role plays, relaxation training, exposures, cognitive restructuring, and reinforcement (Kendall & Hedtke, 2006)

- ☐ Supported components of adolescent depression include psychoeducation, emotional regulation, enhanced social interactions, social skills training, relaxation training, cognitive restructuring, and problem-solving training (Kendall & Hedtke)
- ☐ Child referred to school counselor or psychologist for counseling and assessment if experiences a cardiac event during the school year (Ex: fainting spell or sudden cardiac arrest)
- ☐ Child referred to school counselor or psychologist for counseling and assessment if shows signs of psychological distress, such as anxiety or depression (Ex: increased irritability, sadness, anger outbursts, mood swings, tearfulness, and thoughts of hopelessness or suicide)

5. SCHOOL-BASED TRAININGS

- ☐ All staff (if possible) receives LQTS training in 504 Plan
- ☐ Training topics include LQTS facts, challenges, triggers, symptom displays, identifying a medical emergency, emergency plans, treatment regimens, the use of automatic external defibrillators (AED), cardiopulmonary resuscitation (CPR) and first aid training, treatment, and medication side-effects
- ☐ Medical specialist in LQTS involved in training
- ☐ At least two staff per building (optimally two employees per floor) have current CPR and first aid training through a nationally recognized training organization

- ☐ LQTS resources and references provided
- ☐ Trainers available for questions or follow-up trainings throughout the year

6. EMERGENCY RESPONSE PLANS

- ☐ School requests staff to volunteer to care for adolescent with LQTS
- ☐ Crisis response team established with designated roles (Who alerts crisis response team, removes non-essential people from situation, calls parents and doctor, calls ambulance, administers CPR, gets AED, uses AED, directs emergency personnel in building, briefs emergency responders, handles media, and assists the nurse)
- ☐ Roles assigned to staff for regular school day, after school, and on school trips
- ☐ Specialized training from medical specialist provided (For instance, training should include what to do if a student is unconscious, how contact the nurse, when to call 911, how to contact the student's physician and parents, how to obtain emergency contacts, what if the student's ICD administers shocks, how or when use AED, what to do until ambulance arrives, and how to administer CPR)
- ☐ Emergency plan created or updated with medical specialist/504 Plan committee
- ☐ Emergency response procedures specific to variety of LQTS-specific situations (If faints, pulse not present, unconscious, ICD shocks, and palpitations or dizziness)
- ☐ Emergency response plan written and signed by all parties present in meeting
- ☐ Emergency response plan distributed to all staff responsible for child's well-being

7. DISASTER PREPAREDNESS PLANS

- ☐ Disaster preparedness steps detail how to notify staff, students, and parents of critical emergency/student death (Ex: Secure facts, inform faculty, students, and parents, contact nearby schools, support, resources, debrief, refer high risk students to counseling)
- ☐ Designees appointed (Who secures facts, notifies crisis response team, informs students and staff, provides support, obtains funeral info, and manages media)
- ☐ Plan is written and disseminated to all staff

Appendix B

LQTS Assessment and Interview Guidelines for 504 Planning

Note: The ultimate goal of assessments and interviews are to guide the creation of appropriate LQTS accommodation and intervention plans, helping the schools aim to improve the child's psychosocial functioning and safety.

Assessment Characteristics:

- ☐ Schools should conduct a biopsychosocial assessment: Includes the student's medical, social, educational, and emotional needs (Power & Parrish, 1995)
- ☐ Schools can conduct informal assessment interviews or administer formal standardized measures to evaluate the child's psychosocial, medical, and educational functioning
- ☐ Areas of importance may include physical health history, quality of life, development, cognitive functioning, academic achievement, psychological coping, medical adherence, and family functioning (Rodrigue, Gonzalez-Peralta, & Langham, 2004)
- ☐ Other sources of evaluation: Teacher observations of peer relationships, previous school medical and educational records, psychological test results, child interviews, parent interviews, teacher interviews, medical

professional interviews, medical record reviews, child-self report rating scales, teacher rating scales, and parent rating scales (Clay, 2004)

- ☐ Assessments are often conducted as a result of a 504 request
- ☐ Assessments should be conducted on an ongoing basis, as determined by the intervention team, and should include diverse sources of data

General types of assessments:

- ☐ Assessments for anxiety, depression, and health-related coping: Child Depression Inventory (CDI) for depression; Multidimensional Anxiety Scale for Children (MASC) for anxiety; Self-Perception Profile for Adolescents (SPPA) for self-esteem and self-perceptions; Pediatric Quality of Life Inventory (PedsQL) for health-related quality of life (Varni, Seid, & Rode, 1999; Wichstraum, 1995)
- ☐ Checklists of general stress can also be given, including declines in grades, general mood, worries, anger, fatigue (Clay, 2004)

LQTS Student Assessments:

- ☐ Since LQTS is diverse, it is important to conduct a specialized assessment interview, including relevant academic, family, medical, psychosocial, behavioral, social, and cultural history

- ☐ Ask about the reasons for the assessment, problems they may be having associated with LQTS, the contexts in which the problems are occurring, coping strategies they use, and possible solutions (Clay, 2004)
- ☐ Questions of particular importance include perceptions of social restrictions, co curricular involvement and interest, type of LQTS, triggers, fears, medical events, medications, previous school accommodations, and prior medical and psychological treatments
- ☐ Questions related to self-esteem (SE), should pertain to feelings of social, athletic, and overall competence
- ☐ Questions assessing SE should address the student's thoughts, feelings, and behaviors related to their feelings of self-worth and self-satisfaction (Ex: ask if they believe they are as good in sports as they would like to be. If not, a follow-up question could ask if they attribute their lack in athletics to their personal skill-sets or due to LQTS)
- ☐ To assess social comparisons, one could ask how they view themselves as compared to others their age (their ability to make and keep friends, connectedness to school, opportunities for school activities, involvement in social activities, the number of friends they have, and their opportunities for the future)
- ☐ Ask questions pertaining to the impact of LQTS on school adjustment (Ex: asking how students and staff treat them, followed by inquiring if their

positive or negative interactions are attributed to situational or dispositional characteristics, such as having LQTS)

- ☐ To assess school connectedness and satisfaction, can ask if they have enough extracurricular and social activity opportunities
- ☐ Ask about their knowledge about and perceptions of LQTS (Ex: if unaware that they could die from the condition, it will be critical for school staff to describe LQTS consistent with the family's description)
- ☐ To assess illness-related fears and feelings of vulnerability, schools can ask if they have witnessed a cardiac event before, if they have been symptomatic in front of their peers, the impact of that incident, and if they know others who have LQTS and their outcomes
- ☐ To get an understanding of the child's circumstance and beliefs, one can ask when they were first diagnosed, if they had ever had a symptomatic episode, the circumstances of the event, and the frequency of their medical events
- ☐ Assessment interviews should also have strength-based components, including the student's supports, positive coping, personal assets, sources of strength, and close family contacts

LQTS Parent Assessments:

- ☐ Parents are a critical source of information: Can provide perceptions of the adolescent's functioning
- ☐ Schools should question parents about their concerns regarding their child's health and well-being in school
- ☐ Differences between parent's views and their child's should be discussed openly
- ☐ Health-related questions: How often and what type of medications the child takes, if the school needs to administer medications, if the child experiences medication side-effects, if they typically adhere to the regimen, accommodations they need, if they wear a medical identifier
- ☐ Questions to guide interventions: If the child has food restrictions, such as caffeine, if they are compliant, if they need breaks from physical activities, when resting is important, what accommodations have worked in the past, if they have a pacemaker or implantable cardioverter defibrillator (ICD), if the child has any other known medical or mental health problems, if they have preferred medical contacts, and if they have any other academic or school-related problems
- ☐ It can be helpful to ask parents about their beliefs regarding the LQTS and their family's coping styles as those factors may influence the amount of school-based support that they will seek (Carlson et al., 2004)

- ☐ It is important to inquire about what they feel comfortable disclosing to other students and staff (Clay, 2004).

LQTS Medical Professional Assessments:

- ☐ Medical and mental health professionals, knowledgeable about LQTS, can collaborate in the development of the intervention plan
- ☐ Collaboration can help brainstorm emergency plans, identify and prioritize the most critical needs, enhance problem-solving, and provide medical expertise (Sheridan & Cowan)
- ☐ Schools can ask medical professionals (Ex: cardiologist or the child's primary care doctor) unanswered medical questions, how LQTS affects the child's functioning, what curriculum must be modified, what emergency devices should be purchased, the steps for a crisis plan, and a list of preferred doctors, medical professionals, or hospitals in case of an emergency
- ☐ The child's doctor should be asked about physical activity restrictions, medication management plans, explain the roles of various doctors that care for the child, and help make clear what LQTS is and how it is treated
- ☐ Licensed psychologists who have expertise in adolescent chronic illness can also advocate for the child's social, psychological, and emotional needs

- ☐ Medical and mental health professionals can be asked for resources, availability for staff training, help with equipment training, and assistance in crisis management

Appendix C

Model 504 Accommodation Plan for Students with LQTS

Your School District

School Year: 2009-2010

Date of Report: 6/14/09

This evaluation will be addressing eligibility under Chapter 15, Section 504 of the Rehabilitation Act of 1973. These accommodations will be valid for one calendar year and should be reviewed at the beginning of each new school year. If there are any changes in the student's needs, this plan should be revised as soon as possible.

(THIS IS A CONFIDENTIAL REPORT. RELEASE OF THIS INFORMATION TO ANYONE OTHER THAN THE SCHOOL REQUIRES PARENT SIGNATURE.)

Note: This model 504 Plan lists a broad range of services that may be needed for a child with LQTS. This is not a comprehensive list of services nor will all services be necessary for each child. Each plan must be individually tailored to the unique needs of each student, and multidisciplinary collaboration is encouraged.

Student Name: Adolescent with LQTS

Parent/Guardian: Parent's Name

School: Your School

Birth date: 8/8/1998

Grade: 7

1. REASON(S) FOR REFERRAL AND OBJECTIVES:

Long QT Syndrome (LQTS) is an inherited genetic cardiac disorder, characterized by lengthened time between heartbeats (Arnestad et al., 2007; Bonn, 1997). This electrical cardiac instability places LQTS patients at increased risk for irregular heart rhythms and sudden cardiac arrest (Napolito, Bloise, & Priori, 2006). The harmful effects of LQTS can be triggered by physical activity, such as swimming, sudden auditory sounds (loud noises from alarm clocks, telephones, or doorbells), emotional stress, fright, anger, and sleep (Collins & Van Hare, 2006; Kapetanopolous et al., 2006). The disorder has also been linked to sudden death in young athletes due to the participation in competitive and rigorous athletics (Corrado, 2005). Individuals with LQTS are often restricted from engaging in activities that could lead to heart rate increases. Therefore, LQTS limitations may present obstacles to engaging in athletics and school functions, making school-based services and crisis plans important. The goal of this plan is to provide LQTS services needed to maintain student health and safety, and to respond appropriately to medical emergencies.

2. JUSTIFICATION FOR SERVICES:

Since LQTS is a chronic health condition that places the student at increased risk for abnormal heart rhythms and sudden cardiac arrest, he/she is eligible for a school-based health plan to accommodate his/her medical and school-based needs (Napolito, Bloise, & Priori, 2006).

3. RECOMMENDATIONS/ACCOMMODATIONS

The following related aides, services, or accommodations have been agreed upon by the school district and the parents in addressing the student's educational needs:

1. The gym teachers will provide the student with alternate non-competitive and recreational gym class activities to allow them to participate with the other students as much as possible (as determined by medical professionals, such as allowing them to keep score of games, offering the child to referee or coach, or permitting them to take roll for class)
2. Coaches and after-school supervisors will make revisions to extracurricular activity requirements to allow the student to participate in activities (as approved by medical professionals)
3. If requested or preferred by the student or their family, the gym teachers would permit the student to change for gym in the nurse's office if they have scarring from surgery
4. If needed, teachers will allow the student to leave during class to take

medications

5. If requested, teachers and the school nurse will allow the student to take their medications during breaks, as opposed to leaving during class (Power et al., 2003)
6. All faculty will permit the student to take breaks from physical activity
7. All faculty will allow the student to cease physical activity at any time if they deem it necessary
8. Gym teachers will provide supervision at all times during physical activity
9. A peer buddy system will be developed, as requested
10. The nurse and the 504 Planning team will provide yearly staff trainings
11. The nurse and the collaborative 504 Planning team will develop an emergency response health plan
12. The building administrator will purchase at least one AED and ensure that it is located in an accessible location and operating properly
13. The school counselor will provide support groups and psychological counseling as needed
14. All staff will communicate health-related concerns or 504 Plan breaches with the school nurse who will immediately contact the student's parents, such as if the child was noncompliant with their gym restrictions or missed medications
15. The nurse and counselor will maintain ongoing communication with parents and medical specialists
16. The school nurse will monitor vital signs and observe student responsiveness as needed

17. The school nurse will monitor fluid intake to maintain hydration in hot climates and temperatures
18. The school nurse will monitor food intake to ensure that the child sustains their energy and is able to take their medicine, such as allowing the child to eat snacks during the day
19. The school nurse will monitor restrictions to foods or substances which increase sympathetic arousal, such as those containing caffeine or chocolate
20. School administrators will arrange for nurses to be available during school, after school, and on school trips
21. For field trips, an AED, a designee trained in CPR, AED use, and first aid will be available, along with the student's emergency contact information, an emergency plan, and a communication device, such as a cell phone. **MODEL EXAMPLE-** The designee will alert the nurse and call the school principal, the nurse will administer CPR, the designee will get the AED, the school nurse will use the AED, the designee will call for an ambulance (911), the designee will remove other students and non-essential people from the situation, the nurse will call the student's parents and medical doctor, the designee will direct emergency personnel once they are in the area and brief emergency responders, and the designee will assist the nurse
22. For medical adherence encouragement, the school nurse will assess barriers to treatment adherence monthly, such as discussing with the adolescent their difficulties and successes in abiding by gym-class restrictions (Phelps, 2006)
23. The school nurse will assist the student with creating a chart that tracks their

adherence, and will be encouraged to put prompts, such as daily reminders, in their school calendar

24. A medical management plan will be given to all staff by the school administrator, including the student's name, medical condition, triggers, medications taken, emergency contacts, physician's name and contact information, where the AED is located, and steps in the event of an emergency
25. The school nurse or the student's parents (as determined in the collaborative meeting) will contact local hospitals and emergency health facilities to notify them of the student's condition in the event of a crisis
26. A school reentry program will be established, in the event of an illness-related absence (including ongoing contact with the family, homebound instruction if needed, an extra set of textbooks, curriculum modified as needed, and informing parents about the child's progress on a regular basis)
27. Counseling and psychological services will be offered by the school counselor and school psychologist, as needed
28. Yearly staff training will be provided by the administration, the 504 Plan committee, and a LQTS medical specialist. The training will include instructions about challenges associated with the syndrome, strategies for dealing with them, how to identify a medical emergency, and who to contact for questions or in the event of an emergency. All school employees (if logistically possible based upon each individual school) will be educated about LQTS, emergency plans, treatment regimens, the use of AED's, training in cardiopulmonary resuscitation (CPR) and first aid, implanted cardioverter

defibrillators (ICD), symptom displays, treatment, and possible medication side-effects

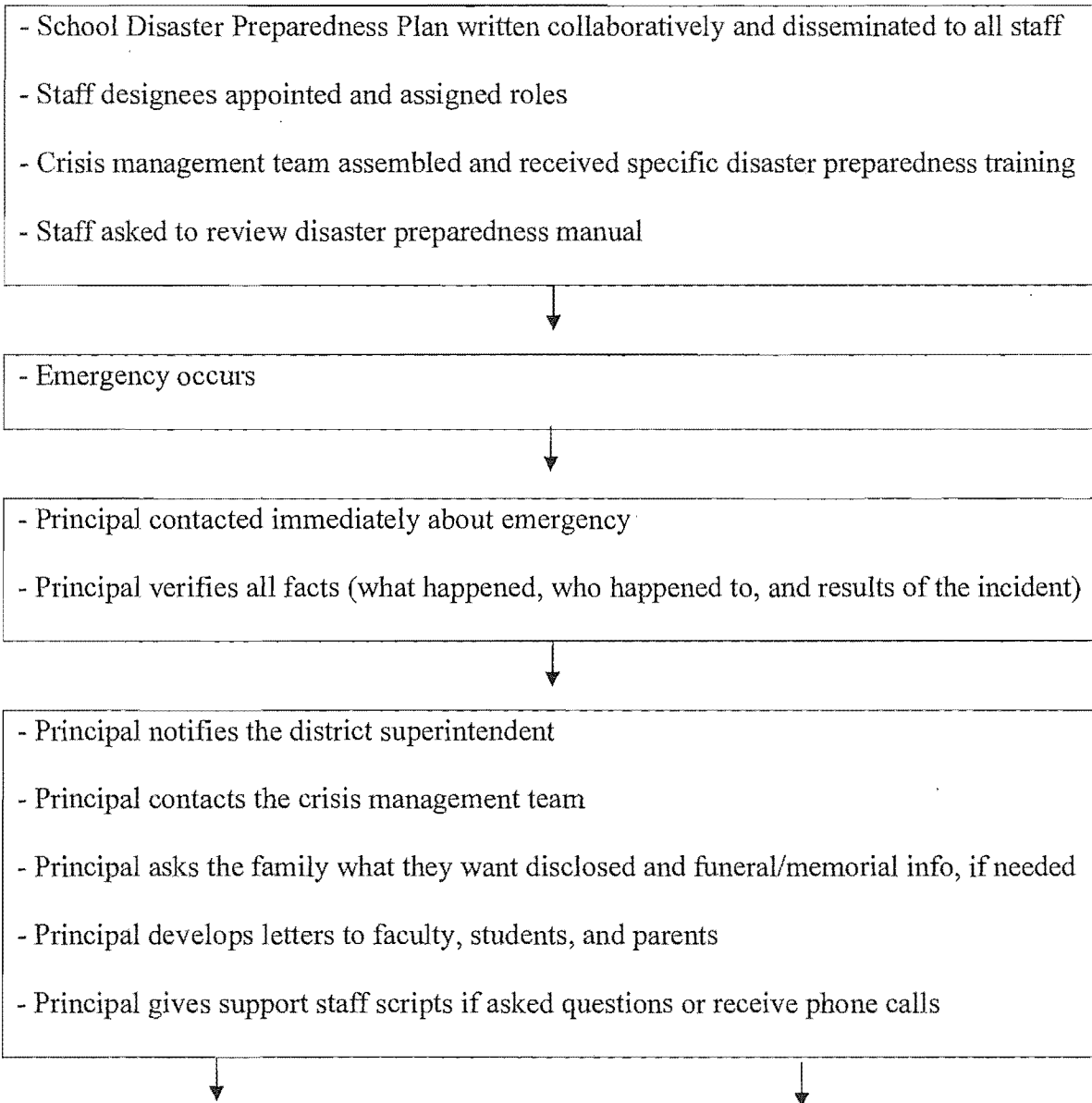
29. With the family's request, provide a picture of the student to approved staff, while ensuring that confidentiality is maintained from other students

4. CRISIS PLAN

1. A district crisis response team is formed, comprised of the school's principal, school counselors, school psychologist, school nurse, and teachers trained in crisis management. These roles and emergency steps are delineated, within the district's emergency manual
2. Conducted with a medical specialist, these individuals will receive additional specialized training (what to do if the student is unconscious, how to contact the school nurse, when to call 911, how to contact the student's physician and parents, steps to obtain emergency contacts, what to do if the student's ICD administers a shock, how to use the AED, when to use the AED, and what to do until the ambulance arrives)
3. Members of the crisis team will be assigned specific roles: **MODEL**
EXAMPLE- The principal will coordinate and alert the crisis team and nurse, the gym teacher will administer CPR, the principal will get the AED, the school nurse will use the AED, the nurse will call for an ambulance (911), the nurse will call the student's parents and medical doctor, the school counselor will remove other students and non-essential people from the situation, the school secretary will direct emergency personnel once they are in the building

Appendix D

Sample School-Based Disaster Preparedness Plan Procedures



- Principal initiates emergency phone chain **if crisis occurs after school hours** (informs all staff of incident including support staff)

- Crisis response designees contact student's parents, and speak personally with teachers throughout the day **if crisis occurs during school hours** (distributes memos with facts, plans for disclosure, changes in daily schedule, funeral information if needed, and staff support offered)



- Principal requests all staff attend a mandatory faculty meeting the *next morning* **if crisis occurs after school hours** (provides facts, plans for disclosure, changes in daily schedule, funeral information if needed, and staff support is offered)



- Principal requests all staff attend a mandatory faculty meeting at the *end of the day* **if crisis occurs during school hours** (additional facts, plans for disclosure, changes in daily schedule, funeral information if needed, and staff support is offered)



- Crisis management team determines if relatives attend any schools within the district
- Crisis management team contacts those principals and counselors
- Crisis management team contacts neighboring school districts for assistance
- Principal determines if substitute teachers needed to relieve classroom teachers



- Principal provides scripts to teachers (consistent info to all students, at the same time)
- Teachers read scripts to students to inform them of incident and dispel rumors (facts, normalization of reactions, how to approach media, and opportunities for counseling)
- Students encouraged to talk about the situation with staff



- Guidance, psychologist, and crisis team offer group and individual counseling
- Guidance provides students, parents, and faculty with resources and information regarding reactions to crisis or grief, and psychological services



- Staff observe vulnerable students and refer to guidance and/or crisis management team for additional psychological services



- Principal distributes memo to parents (facts, available student psychological services, how to address the media, and funeral information if needed)
- Memo is mailed or given to students to bring home



- Principal, guidance, and crisis team provide debriefing a few days after the incident (situation processed with students and staff; funeral/memorial info provided if needed)
- Staff asked to refer high risk students to the guidance and crisis management team for next few months
- Continued student and staff discussions encouraged



- Principal, guidance, and the crisis management team meet at the end of the first day after the incident was disclosed to determine if school needs additional crisis services

¹ This flow chart was modified and adapted from a disaster preparedness protocol developed at the Avon Grove School district in West Grove, PA.

Appendix E

Sample Announcement to Staff

Dear staff,

It is with great sadness to announce that (student's name), a student in our school, (event that occurred). Please extend your warm wishes to their family and friends during this time of need. At this time, (funeral arrangement information, if needed).

- Event:**
1. What happened? (Facts of the event)
 2. When did it happen?
 3. Plans for disclosure
 4. Changes in daily schedule
 5. Staff support offered

- Funeral info:**
1. Where and when
 2. *If no information is available* say, "At this time, funeral arrangements are incomplete. When service arrangements have been confirmed, you will be notified."

Teachers will receive a memo to read to their students with an appropriate announcement concerning this tragic event. Support staff will receive a script in the event that they are questioned or receive phone calls. The crisis response team including guidance counselors, administration and trained staff are available to offer support to students and faculty/staff.

If approached by the media, please do not make any comments. Our district's media designee will be providing any necessary details.

Sincerely,

Principal

² This announcement to teachers was adapted from a disaster preparedness protocol developed at the Avon Grove School district in West Grove, PA.

Appendix F

Sample Announcement to Students

TO: Staff

RE: Announcement to Students

DATE: 6/14/2009

**** IF YOU NEED ASSISTANCE IN READING THIS ANNOUNCEMENT TO STUDENTS, PLEASE CONTACT ADMINISTRATION OR GUIDANCE ****

- Read this statement exactly as written:

We would like to announce that (student's name), a student in our school, (event that occurred). Please extend your warm wishes to their family and friends during this time of need. At this time, (funeral arrangement information, if needed).

- Event:**
1. What happened? (Facts of the event)
 2. When did it happen?
 3. Plans for disclosure
 4. Changes in daily schedule
 5. Student support offered

Funeral info: 1. Where and when

2. *If no information is available* say, “At this time, funeral arrangements are incomplete. When service arrangements have been confirmed, you will be notified.”

Our school’s crisis response team including counselors, administration, and trained staff will be available students needing support in dealing with this event. Your teachers will advise you of the location and times available this support.

**Please allow students to contact any of our crisis team members if they have been affected by this news: (List team members at each school)*

****** If the crisis occurs during non-school hours, a mandatory faculty meeting will be held the next morning, where the formal “announcement to students” will be distributed to faculty. (All administrators and guidance counselors will be informed of the crisis situation via phone chain prior to the next school day).

****** If the crisis occurs during school hours, a formal “announcement to students” will be distributed to staff, and a mandatory faculty meeting will be held at the end of that day.

³ This announcement to students was adapted from a disaster preparedness protocol developed at the Avon Grove School district in West Grove, PA.

Appendix G

Sample Announcement to Parents

Dear Parent/Guardian,

It is with great sadness to announce that (student's name), a student in our school, (event that occurred). Please extend your warm wishes to their family and friends during this time of need. At this time, (funeral arrangement information, if needed).

Event:

1. What happened? (Facts of the event)
2. When did it happen?
3. Student support offered

Funeral info:

1. Where and when
2. *If no information is available* say, "At this time, funeral arrangements are incomplete. When service arrangements have been confirmed, you will be notified."

If approached by the media, please discourage all children from disclosing sensitive information. If you have any questions concerning your child's reaction to this situation, including signs and symptoms of grief, please contact the school.

Sincerely,

Principal

⁴ This announcement to parents was adapted from a disaster preparedness protocol developed at the Avon Grove School district in West Grove, PA.