LONG QT SYNDROME: HELPING NURSE PRACTITIONERS SCREEN, DIAGNOSE AND MANAGE IN PRIMARY CARE

By

SHARLA K. PETERSON, RN, BSN

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The members of the Committee appointed to examine the clinical project by SHARLA K. PETERSON find it satisfactory and recommend that it be accepted.

Chair: Louise Kaplan, PhD, ARNP

Jaekie Barasik, PhD, ARNP

Naomi Lungstrom, MN, ARNP
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Abstract

By Sharla K. Peterson, RN, BSN
Washington State University
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Chair: Louise Kaplan

Sudden and unexpected cardiac death in children and young adults are tragic events often related to a disturbance of the heart's electrical conduction system, resulting in lethal arrhythmias. Unfortunately, the underlying conditions leading to sudden cardiac death (SCD) are often not suspected or are misdiagnosed. One of these conditions is Long QT Syndrome (LQTS). LQTS is a pathological cause of loss of consciousness, syncopal episodes and seizure-like activity but is often not recognized or accurately diagnosed prior to a sudden death or significant cardiac event. This article presents a case report of a young woman with LQTS who experienced an aborted SCD event. The article also provides a review of LQTS and recommendations for nurse practitioners regarding screening, management and the education of patients and families.
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Long QT Syndrome

Long QT Syndrome Introduction

Sudden and unexpected cardiac death in children and young adults are tragic events often related to a disturbance of the heart’s electrical conduction system resulting in lethal arrhythmias. Unfortunately, the underlying conditions leading to sudden cardiac death (SCD) are often not suspected or are misdiagnosed by primary healthcare providers and consequently go untreated. One of these conditions is Long QT Syndrome.

Long QT Syndrome (LQTS) is a pathological cause of loss of consciousness and syncopal episodes but is often not recognized or accurately diagnosed prior to a sudden death or significant cardiac event (Trusty, Beinborn, & Jahangir, 2004). LQTS is a frequently missed diagnosis both in primary and acute care settings. This article presents a case report of a young woman with LQTS who experienced an aborted SCD event. The article also provides a review of LQTS and recommendations for nurse practitioners (NPs) regarding screening, management and the education of patients and families.

Case Report

Early one morning in 1996, Christina, age ten, got up to turn off her alarm when she had her first fainting episode. Her parents heard a thump and found Christina on the floor disoriented and with no recollection of what had happened. Two months later, she had a similar episode. An EEG to evaluate for seizures was done and was found to be normal. Approximately a year later, her family heard her fall, she was found on the floor, moaning loudly, stiff and unresponsive with glazed eyes, tonic clonic movement and loss of bladder control. She had stopped breathing for a few seconds. Christina had two more episodes over the next several months. In 1998, she was started on carbamazepine for seizures despite a second negative EEG.
After two years with no “seizures,” Christina’s doctor weaned her off of the carbamazepine. While on vacation, she had another “seizure” just after her mother took a group photo with a flash. The carbamazepine was restarted. Another two years went by and Christina seemed to be under control. One morning when the alarm clock went off, Christina had another “seizure,” during which she was gasping for air and stopped breathing. Her dose of carbamazepine was increased. A month later, after a similar event, her dose of carbamazepine was increased and topiramate was added.

Christina continued to have “seizures” despite repeated increases in carbamazepine and topiramate doses. The topiramate was replaced with zonisamide, however, the “seizures” continued. She reported feeling at times like the air was being sucked out of her before she would faint and seize. In 2003, while standing in front of her class, she fell to the floor, went unconscious, stopped breathing for several seconds, turned blue and started gasping. Christina was then hospitalized for an extensive seizure work up. Despite no evidence of seizures, she continued to be managed with anti-epileptic drugs. She continued to have episodes in which she would lose consciousness. Some of these events would occur after being woken up, answering the doorbell, or after a telephone rang. She also reported her heart would beat hard and fast and that she had difficulty breathing.

In November 2005, now aged twenty, Christina’s life took a tragic turn. While sleeping, she started to have one of her “seizures.” Christina’s husband was woken by her moaning, kicking and gasping for air. She stopped breathing. Unable to arouse her and finding no carotid pulse, he called 911. The EMTs found Christina in fine ventricular fibrillation. Three attempts with defibrillation were required before she returned to a sinus rhythm. Christina was in the shock-trauma unit for over three weeks in a coma and intubated for 2 weeks. She was finally
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diagnosed as having LQTS. She had, however, suffered an anoxic brain injury and stroke resulting in impairment in her short-term memory, balance, and speech. She was genetically tested and found to have a genetic mutation associated with LQTS.

Prior to Christina’s sudden cardiac death, no electrocardiogram (ECG) was ever obtained. Her medical care was solely directed at seizure management despite having negative seizure workups. Could this tragic event have been prevented if a more thorough and detailed history of events, family history and physical exam had been completed? Could this event have been prevented if an ECG was performed during the initial evaluation or when attempts to control the seizure-like activity failed? The intent of this article is to assist healthcare providers to better identify and manage LQTS and prevent tragic events from occurring.

The Pathophysiology of LQTS

Long QT Syndrome is a genetic disorder of cardiac repolarization resulting from alterations in the transmembrane ionic channel flow of sodium, potassium, and calcium (Fair & Mason, 2004). It is characterized by a prolonged ventricular repolarization phase of the cardiac muscle after the action potential and associated ventricular arrhythmias, syncope and sudden death (Fair & Mason, 2004). LQTS is identified on ECG by a prolonged QT interval (Moss, 2003).

QT refers to the interval between the beginning of depolarization through the end of repolarization of the cardiac cycle (Fair & Mason, 2004). The QT interval is measured from the onset of the Q wave to the end of the T wave (Moss, 2003). Normal QT intervals should measure less than 0.44 seconds (Moss, 2003). See Figure 1 for an example of a normal ECG. Generally, leads II, V3, and V4 are the best leads to use in measuring the QT interval (Moss, 2003). Figures
2 through 4 are examples of Christina's electrocardiograms, which all demonstrate a prolonged QT interval.

As the QT interval lengthens, the likelihood of abnormal electrical depolarization during the QT interval increases (Ashworth, Levsky, Marley, & Kang, 2005). If such an event does occur, an arrhythmia may be generated, in particular the ventricular tachycardia Torsades de Pointes (TdP). LQTS patients carry a relatively high propensity of developing TdP (Beery, Shooner, & Benson, 2007). Episodes of TdP are often self-terminating, but have the potential of deteriorating into lethal ventricular fibrillation leading to sudden death (Wever & Robles de Medina, 2004).

Long QT Syndrome can also occur in an acquired form due to certain medications. Current information on drugs known to cause the acquired form of LQTS can be found on http://long-qt-syndrome.com/lqts_drugs.html or http://www.qtdrugs.org/. See Table 1 for a list of the common medications. The acquired form is not covered in this article.

The Electrophysiology of LQTS

Normally, ventricular repolarization time decreases with faster heart rates or increases with slower heart rates, causing the QT interval to vary. An uncorrected measurement may not provide an accurate reflection of the QT interval. The QT correction (QTc) is the QT interval that is corrected for heart rate and is calculated with Bazett's formula, QTc=QT√RR with all intervals measured in seconds (Drew et al., 2005). The value of RR in Bazett’s formula is the measured distance from the previous R wave to the following R wave. A normal QT corrected for heart rate is less than 0.45 seconds for men and less than 0.46 seconds for women (Drew et al., 2005). See Figure 5 for an example of implementing ECG measurements into Bazett’s
formula. Today's standard 12-lead ECG will often provide both the corrected and uncorrected QT intervals. Assessing the QT interval may help avoid missing the diagnosis.

In addition to assessing the QT interval, it is necessary to review the ECG tracing for T wave abnormality. A diagnostic feature of LQTS is T wave alternans, a beat-to-beat variation in the T wave amplitude and a sign of enhanced electrical instability (Fair & Mason, 2004). Additional abnormal T wave diagnostic patterns include bifid (double hump), low amplitude bifid and late onset (Leroy & Russell, 2004). See Figure 6 for examples of T wave abnormalities.

Genetic Aspects of LQTS

Genetic research indicates that there are six genes responsible for LQTS with 90% of the genetically confirmed diagnoses being genes LQT1 and LQT2 (Beery, Shooner, & Benson, 2007). The remaining 10% are accounted for by genes LQT3, LQT4, LQT5, and LQT6 (Wever & Robles de Medina, 2004). An estimated 1 case per 5,000-10,000 live births is affected by LQT1 (Ashworth, Levsky, Marley, & Kang, 2005). Cardiac events such as syncope, arrhythmias and SCD tend to occur under specific circumstances in a gene specific manner (Moss, 2003). LQT1 and LQT2 have a higher incidence of cardiac events than the other variations (Wever & Robles de Medina, 2004). In general, LQT1 patients have arrhythmias triggered by vigorous physical stress, activities or exercise, particularly diving and swimming (Wever & Robles de Medina, 2004). LQT2 patients have more arrhythmias associated with acute arousal type emotions (Wever & Robles de Medina, 2004). Arrhythmias can even be triggered by a sudden loud noise, such as the ringing of an alarm clock, telephone, or a doorbell. LQT3 patients experience events without emotional arousal during sleep or at rest (Wever & Robles de Medina, 2004). The probability of experiencing a cardiac event increases significantly during adolescence in all three genotypes (Ashworth, Levsky, Marley, & Kang, 2005).
Scope of the Problem

In the United States, SCD due to LQTS in children is thought to be three times more common than childhood leukemia ("SADS Foundation News," 2006) and is estimated to cause over 4,000 deaths each year (Ashworth, Levsky, Marley, & Kang, 2005). Syncopal episodes are particularly frequent in the 10 to 20 year old age group, higher in males before puberty and increased in adult women (Moss & Robinson, 2002). Females have a significantly greater risk of having fatal cardiac events than males (Sauer et al., 2007).

There are several factors that prevent the true scope of LQTS incidence and prevalence from being fully known. First, it is estimated that about one third of patients are asymptomatic and never perceive a need to seek medical care (Wever & Robles de Medina, 2004). Second, there is a lack of structural heart disease found during diagnostic imaging studies or during autopsy (Leroy & Russell, 2004). Third, there can be subtle or no ECG findings even in patients with a known LQTS diagnosis. Finally, SCD due to LQTS may be the initial and only presentation of LQTS and it may be difficult to ascertain if LQTS was the cause of death (Leroy & Russell, 2004).

Clinical Presentation of LQTS

Individuals with LQTS generally appear to be in overall good health with no obvious structural or mechanical abnormalities of their hearts. There are no physical abnormalities to detect with LQTS (Robinson & Allan, 1998). Signs and symptoms associated with congenital LQTS generally begin during childhood or adolescent years (Viskin, 1999). In 30% to 40% of children and adolescents, however, their first manifestation of the disorder may be a syncopal episode or seizure-like activity caused by a transient rapid heart rhythm that was triggered by emotional or physical stress (Vetter, 2007). Cardiac arrest may be the initial presentation,
especially in infants (Viskin, 1999). Arrhythmias may also occur while an individual sleeps causing nightmares, seizure-like episodes or nocturnal death (Fair & Mason, 2004).

The syncope caused by a life threatening cardiac arrhythmia in LQTS is sudden, usually without any type of warning (Moss & Robinson, 2002). It often occurs during or immediately following exercise, emotional stress, being startled or hearing certain noises. The syncopal episode may be accompanied by gasping or apnea, absence of a pulse or a rapid irregular pulse and in some instances the patient may become cyanotic (Moss & Robinson, 2002). Loss of bladder control and seizure-like activity may also occur. Usually the loss of consciousness lasts from one to several minutes and the individual generally recovers within a few minutes (Ashworth, Levsky, Marley, & Kang, 2005).

Fainting episodes may be a warning sign of a potentially life threatening arrhythmia and the forerunner of sudden death. Healthcare providers often misdiagnose LQTS in children and adolescents after the first episode of syncope or seizure-like activity. A seizure disorder is the most common misdiagnosis (Bell & Kozak, 1996), (Viskin, 1999) & (Wever & Robles de Medina, 2004). The seizure-like activity is most likely due to cerebral hypoxia caused by arrhythmias that decrease cerebral perfusion. A syncopal episode or seizure-like activity in a young, otherwise healthy individual requires a full medical examination including an ECG to evaluate the QT interval.

**Screening in Primary Care**

Screening for this disorder is difficult. The most prudent, effective, and cost efficient method of screening for LQTS is thorough history taking, physical examination and use of ECGs with accurate interpretation (Ashworth, Levsky, Marley, & Kang, 2005). Table 2 is a form that can guide the screening. Approximately two-thirds of patients are diagnosed with LQTS with
routine ECG screening or because a family member is diagnosed. One-quarter of people with LQTS are asymptomatic at the time of diagnosis and may remain asymptomatic throughout their lives (Leroy & Russell, 2004).

Nurse practitioners face the challenge of determining who and when to screen for LQTS. The initial challenge in primary prevention is to identify individuals who are at greatest risk before a tragic event occurs (Obias-Manno & Wijetunga, 2004). NPs should consider screening for LQTS as part of their differential diagnosis in an otherwise healthy patient with the following problems.

1. Report of loss of consciousness, syncope, near syncopal episodes or seizure-like activity during or immediately following exercise, emotional stress/excitement, being startled or after hearing certain noises. Patients with a history of a syncopal event have an increased risk of experiencing recurrent events (Moss, 2003).

2. Chest pain that is unusual or consistent despite activity level, palpitations and/or excessive shortness of breath during exercise. Individuals resuscitated from a prior cardiac arrest have a higher risk of subsequent fatal events (Moss, 2003).

3. A family history of unexplained sudden death in otherwise healthy family members, unexplained death during swimming or while driving a car, syncope, death during "seizures" or a family history of "seizure" disorders with no diagnostic EEGs. A positive or potential family history of LQTS increases the likelihood of LQTS in an individual especially when the patient is symptomatic (Leroy & Russell, 2004). The absence of a family history of these events, however, does not exclude the diagnosis given that one-third of patients diagnosed with LQTS have a negative family history (Beery, Shooner, & Benson, 2007).
Obtaining a Complete History

Confusion often arises if a superficial rather than comprehensive history is obtained when there is complaint of palpitations, chest pain, syncope, or seizures. Approximately one-third of patients with LQTS are diagnosed during these types of health evaluations (Leroy & Russell, 2004). In 9% of patients younger than 21 years, the presenting problem is SCD (Leroy & Russell, 2004). The most frequent ventricular arrhythmia associated with LQTS, TdP, is similar in presentation to seizures or benign syncope. It is imperative to differentiate between LQTS associated arrhythmias, syncope, and seizures. While important, syncope and seizures do not generally lead to SCD. The NP should develop a set of differential diagnoses while maintaining a high index of suspicion of LQTS when evaluating seizures, palpitations, or recurrent syncopal episodes in a patient (Vetter, 2007). Many of the potential causes of syncope, palpitations, or seizure like activity can be discarded from the differential diagnoses during the exam.

Sports Pre-Participation Screening

The American Heart Association (AHA) and the American College of Cardiology (ACC) promote sports pre-participation screening to insure the health and safety of athletes (Trusty, Beinborn, & Jahangir, 2004). The AHA and the ACC developed guidelines to identify cardiovascular conditions that predispose to SCD allowing earlier intervention by healthcare providers, families, and coaches. The groups recommend that all high school and college athletes have a cardiovascular screening performed before they participate in organized sports with physical re-evaluation every 2 years and an interim history in the alternate years. The AHA and the ACC also recommend the screenings should include a detailed personal and family history and physical examination. ECGs should be performed on athletes with signs or symptoms suggestive of cardiovascular conditions (Trusty, Beinborn, & Jahangir, 2004). Symptoms that the
athlete may consider irrelevant may indicate a life threatening condition. Approximately 64% to 78% of cardiac related conditions that may either prohibit or modify sports participation can be determined by either a history of cardiovascular disease or a family history of SCD (Trusty, Beinborn, & Jahangir, 2004).

Diagnostic Testing with ECGs

There are no physical abnormalities to detect in LQTS. Physical examination provides no relevant data, however, physical findings may correlate with other differential diagnoses that lead the NP to eliminate LQTS as the cause of a symptomatic individual’s problem. It is essential to assess for electrical disturbances found on ECG during the depolarization through the repolarization phase. These findings can be subtle and easily overlooked or misinterpreted (Leroy & Russell, 2004). Patients presenting with complaints of dizziness, palpitations, near syncope or syncope, new onset seizure or aborted cardiac arrest should have an ECG to identify arrhythmias and a prolonged QT interval (Fair & Mason, 2004). This should be done regardless of the patient’s age. ECG is the most readily available, cost effective, and least invasive means of testing for LQTS (Trusty, Beinborn, & Jahangir, 2004). If the findings are inconclusive with a resting twelve lead ECG, a stress ECG should be ordered. It monitors for a prolonged QT interval despite the increased heart rate as normally the QT interval shortens during exercise (Trusty, Beinborn, & Jahangir, 2004). Holter monitors can also be useful for monitoring QT intervals over time and in a variety of daily activities.

ECG Diagnostic Criteria

The standard diagnostic criteria for LQTS is marked QT prolongation in which the QTc is >0.45 seconds for men and >0.46 seconds for women (Wever & Robles de Medina, 2004). Other ECG diagnostic criteria include abnormal ST wave contours, macroscopic T wave
alternans, lower than normal heart rates and the occurrence of sinus node pauses (Wever & Robles de Medina, 2004). Marked QT prolongation of greater than 0.47 seconds alone is diagnostic even if there are no other ECG abnormalities. When the ECG reveals a less impressive QT prolongation, other factors such as positive family history, occurrence of symptoms associated with LQTS or a documented TdP should be taken into account to aid the NP with diagnosis and increase the likelihood of a clinically accurate diagnosis (Beery, Shooner, & Benson, 2007).

Significance of ECG Findings

The most significant risk factor for cardiac events and SCD is the length of the QTc interval (Moss, 2003). Patients, independent of cardiac function, age, sex, or use of medications known to affect the QT interval, with a prolonged QTc interval greater than 0.44 seconds have a two to three times higher risk of SCD than those with shorter intervals (Fair & Mason, 2004). Ten percent to 50% of adult LQTS patients and 15% of children with LQTS have either a normal or borderline ECG on initial evaluation so a normal QT interval does not exclude the diagnosis (Leroy & Russell, 2004).

Cardiology Referrals

Referral to a cardiologist is indicated for all patients with a prolonged QT interval or abnormal T wave on ECG. Additional referrals should be made for patients with a normal QT interval who report recurrent syncope, syncope occurring with exercise or fright, unexplained seizure and a family history of sudden death (Robinson & Allan, 1998). Cardiology referral is indicated when additional diagnostic tools are needed for more conclusive diagnosis. Consulting with a cardiologist is encouraged whenever the NP needs additional expertise in evaluating a patient’s potential for LQTS when ECG findings are equivocal.
Genetic Testing

A patient diagnosed or highly suspected to have LQTS should have genetic testing performed. Genetic testing is the gold standard for diagnosis (Vetter, 2007). Immediate family members should also be tested if there is a definitive diagnosis of LQTS. Genetic testing can determine carriers of the mutation and asymptomatic family members who are candidates for treatment. However, a negative test does not exclude the diagnosis of LQTS due to 30% of the genes or mutations have yet to be identified (Vetter, 2007).

Treatment for Symptomatic Patients

Effective treatment can be implemented immediately after diagnosis and life-threatening events prevented. The first step in implementing appropriate management of LQTS is recognizing the potential for devastating consequences in the absence of treatment (Fair & Mason, 2004). Cardiac events can be prevented and reduced in both frequency and severity; however, people with LQTS have a poor prognosis if diagnosis and treatment are delayed (Moss, 2003). Symptomatic individuals with treatment are at an annual risk of 5% per year for abrupt syncopal events and a significantly lower mortality rate (Leroy & Russell, 2004). Symptomatic patients without treatment have a mortality rate of 20% in the year following the initial syncopal episode (Bell & Kozak, 1996). Symptomatic patients without treatment also have a 10-year mortality rate that approaches 50% (Leroy & Russell, 2004). These statistics support initiating treatment, which may include pharmacological therapy, invasive treatment and nonpharmacological therapy.

Pharmacological Therapy

Treatment goals for LQTS are targeted at regulating the heart rate, normalizing the QT interval length and preventing early ventricular depolarizations and arrhythmias (Ashworth,
Levsky, Marley, & Kang, 2005). Therapy is life long once begun. Beta-blocking drugs, in particular propranolol, are the treatment of choice for patients with history of syncope, aborted cardiac arrest, and in patients from high-risk families with genetic linked LQTS. Beta-blocker efficacy results from the attenuation of the adrenergic mediated mechanisms that are triggered in LQTS (Moss, 2003). Although they have little or no effect on QTc duration, beta-blocker therapy reduces the frequency of syncopal events (Moss, 2003). Beta-blockers, however, do not provide total protection against fatal cardiac events (Moss, 2003). Treatment with propranolol has shown an impressive relative risk reduction of 41% for syncope and SCD (Bell & Kozak, 1996). NPs should monitor ECGs, vital signs and symptoms after drug therapy is initiated to assess appropriate dosing and efficacy.

Consistent use of medications is paramount for the patient to understand. Once beta-blockers are initiated, continuous use is essential given that life-threatening arrhythmias have a greater likelihood to occur with discontinuance (Moss, 2003). Beta-blockers are generally short acting medications; missing even one dose of the medication may leave the patient unprotected (Moss, 2003). The NP plays an essential role in educating the patient about the medications and stressing their importance.

Women being treated using beta-blockers may have concerns if they wish to become pregnant. There is a small additional risk for cardiac events during pregnancy, however, for up to nine months post partum the risk is significantly higher (Seth et al., 2007). This risk fortunately is significantly reduced with the use of beta-blockers (Seth et al., 2007). Risk to the fetus of a teratogenic defect is extremely small so pregnant patients on beta-blocker therapy should not stop their medications (Bell & Kozak, 1996). Beta-blockers, specifically propranolol, can be safely taken during conception, pregnancy and breast-feeding in young women ("SADS
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Foundation News,” 2006). This information should provide reassurance for female patients with LQTS.

**Invasive Therapy**

Implanted cardioverter defibrillators (ICDs) are not considered a first line treatment for patients with LQTS but are a form of invasive treatment. High-risk patients with a history of aborted SCD, symptoms despite medical therapy, markedly long QT intervals (>0.60 seconds) and family histories of SCD are candidates for ICDs (Leroy & Russell, 2004). Beta-blockers should be also be used in conjunction with ICDs to offer the safest and most effective form of therapy for high-risk patients (Moss, 2003). While the cardiologist will manage the ICD, the NP can also be a resource for questions or concerns the patient may have.

**Nonpharmacological Therapy and Management**

Implementing nonpharmacological management is an important aspect of care for the NP to assist the patient. Once a symptomatic patient is stabilized, routine daily activity is recommended including sexual activities (Trusty, Beinborn, & Jahangir, 2004). Competitive athletics should generally be avoided (Robinson & Allan, 1998). Patients should avoid adrenergic-type stimuli that can trigger life-threatening arrhythmias such as sudden loud noises, doorbells, alarm clocks, rock concerts, fireworks and firearms (Leroy & Russell, 2004).

**Treatment of Asymptomatic Patients**

It is impossible to predict if and when an asymptomatic patient will become symptomatic and require treatment (Fair & Mason, 2004). There are two approaches to treatment of asymptomatic patients. The first approach provides treatment at the time of diagnosis for all asymptomatic patients under the age of 40 (Fair & Mason, 2004). The second approach is to treat asymptomatic patients only if they have high-risk characteristics. These include a QTc interval
15 I. Jong QT Syndrome

>0.60 seconds, T wave alternans or a family member with an unexplained sudden death (Fair & Mason, 2004). These patients should be referred to a cardiologist for initial therapy and management.

Education/Awareness

Significant efforts to increase both healthcare professionals and public awareness of LQTS have been made. Families should be strongly encouraged to take cardiopulmonary resuscitation (CPR) classes and to stay current with these skills. In addition to CPR, the use of automated external defibrillators (AEDs) should be encouraged if they are available. AEDs decrease the time from cardiac arrest to defibrillation, thus improving the survival rate for victims (Ashworth, Levsky, Marley, & Kang, 2005). Families should be reassured that they are capable of successfully performing these life saving measures.

Medic Alert bracelets or necklaces are highly recommended by the Sudden Arrhythmia Death Syndromes Foundation (“SADS Foundation News,” 2006) to help identify patients who have a potentially life threatening form of arrhythmia. Medical personnel are more likely to appropriately treat patients with LQTS and not mistake a cardiac event as benign syncope or a seizure when bracelets are worn. Schools and work places should also be aware of the diagnosis and appropriate interventions. Schools need to be notified of any limitations for participation on team sports and other physical activities.

The Nurse Practitioner’s Role

Following the diagnosis of LQTS, counseling and support are a necessary part of the NP’s role. There is no doubt that the diagnosis of LQTS has a permanent and life-changing impact on the family (Vetter, 2007). Many of the issues a patient with LQTS encounters are emotionally charged. It is paramount the NP develops a warm and trusting relationship with the
patient and the family. Patients may experience complicated emotions and challenges as they cope with and accept their diagnosis. Their stress may be easily exacerbated by fear of another cardiac event occurring. Patients may be faced with the reality of their own mortality after a near fatal event that required defibrillation (Robinson & Allan, 1998). Parents may have feelings of guilt and blame themselves for “giving this to my child” (Leroy & Russell, 2004). Some patients may have a negative attitude or resentment when they are informed that treatment is life long. The possibility an ICD may be needed can generate mixed emotions. Patients may be anxious regarding asymptomatic family members who have not been diagnosed and are not receiving treatment. NPs must be attentive and sensitive to their patients’ individual needs. They should convey a sense of hope and optimism. Referrals for crisis counseling may be appropriate.

The NP is also faced with the challenge of placing and reinforcing limitations on a patient that may significantly impact their activities, hobbies and social interactions. One of the greatest stressors for patients is lifelong restriction from competitive sports (Leroy & Russell, 2004). Patients may be encouraged that once they are stable, they may participate in less intense forms of exercise and activities. The NP should encourage the use of resources and support groups such as those noted in Table 3.

Nurse practitioners are well positioned and educated to be able to screen and identify at-risk individuals, assist in the diagnostic process, initiate appropriate referrals, participate in medical management, educate, provide emotional support to patients and their families and contribute to increasing public awareness.
References


Figure 1 Example of a normal ECG (Moss & Robinson, 2002)
Figure 2 Christina’s ECG

ID: 29-JUL-1985 (20 y)
Female
ID: CA-2005 03:21:04 Hospital
29-JUL-1985 (20 yr) Vent. rate 66 BPM Normal sinus rhythm
Female Caucasian
PR interval 166 ms
QRS duration 92 ms Prolonged QT
Room: QTRTc 556 ms
P-R-T axes 37 113 85
When compared with ECG of 29-NOV-2005 03:28:
Serial changes of Anterior Infarct Present

Technician
Test indication: CHANGES
READING DR/PANEL
Referred by:
Confirmed by:

EDT: 19:38 29-NOV-2005 DH 3CO
25mm/s 19mm/mV 150Hz 005E 128 235 CID: 1
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Long QT Syndrome

**Figure 3 Christina’s ECG**

<table>
<thead>
<tr>
<th>Date</th>
<th>Time</th>
<th>Room</th>
<th>Location</th>
<th>Sex</th>
<th>Race</th>
<th>PR Interval</th>
<th>QT Interval</th>
<th>QRS Duration</th>
<th>P-R-T Axes</th>
<th>ECG Abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>29-JUL-1985 (20 yr)</td>
<td></td>
<td></td>
<td></td>
<td>Female</td>
<td>Caucasian</td>
<td>156 ms</td>
<td>518/559 ms</td>
<td>96 ms</td>
<td>124 108</td>
<td>Abnormal ECG When compared with ECG of 28-NOV-2005 10:12, Lateral Infarct No longer present.</td>
</tr>
</tbody>
</table>

Abnormal ECG

Prolonged QT

Ectopic atrial rhythm Right axis deviation

Possible Right ventricular hypertrophy

T wave abnormality, consider anterolateral ischemia
Figure 4 Christina’s ECG

Normal sinus rhythm
Right axis deviation
Prolonged QTc interval
Right ventricular hypertrophy
Nonspecific T wave abnormality
Abnormal ECG

When compared with ECG of 02-DEC-2005 16:14, no significant change was found.
Figure 5 Implementing ECG measurements into Bazett’s formula (Leroy & Russell, 2004)
Figure 6 Examples of abnormal T waves (Bell & Kozak, 1996)

1. Normal QT interval
2. Wide-based, slowly generated T-wave
3. Wide-based, double hump T-wave
4. Low amplitude deflection on descending limb of T-wave
5. Indistinct termination of T-wave (T-U complex)
6. Sinusoidal, slowly generated T-wave
7. T-wave inscribed after prolonged ST segment
Table 1
List of common medications known to prolong the QT interval (www.sads.org)


*Anti-arrhythmic/Abnormal heart rhythm*
Amiodarone (Pacerone, Cordarone), Disopyramide (Norpace), Dopamine (Intropine), Flecainide (Tambocor), Procainamide (Pronestyl, Procan), Quinidine (Quinaglute, Cardioquin), Sotalol (Betapace)

*Antibiotic*
Azithromycin (Zithromax), Claritromycin (Biaxin), Erythromycin (E.E.S., Erythrocin), Gatifloxacin (Tequin), Levofoxacin (Levaquin), Moxifloxacin (Avelox), Ofloxacin (Flloxin), Pentamidine (NebuPent, Pentam), Sparfloxacin (Zagam), Telithromycin (Ketek)

*Antidepressants*
Venlafaxine (Effexor)

*Anti-emetics*
Dolasetron (Anzemet), Droperidol (Inapsine), Granisetron (Kytril), Ondansetron (Zofran)

*Anti-psychotics*
Chlorpromazine (Thorazine), Clozapine (Clozaril), Haloperidol (Haldol), Lithium (Eskalith, Lithobid), Quetiapine (Seroquel), Risperidone (Risperdal), Thioridazine (Mellaril) Ziprasidone (Geodon)

*Asthma/Bronchodilator*
Albuterol (Ventolin, Proventil), Ephedrine (Broncholate, Rynatuss, Bronkaid, Primatene), Levomethadyl (Xopenex), Metaproterenol (Alupent, Metaprel), Phenylephrine (Neosynephrine), Pseudoephedrine (Pedia care, Sudafed), Salmetrol (Serevent), Terbutaline (Brethine)

*CNS stimulant/ADHD*
Amphetamine/dextroamphetamine (Adderall), Atomoxetine (Strattera), Dextroamphetamine (Dexadrine), Methylphenidate (Ritalin, Concerta)

*Muscle Relaxant*
Tizanidine (Zanaflex)

*Opiate agonist/Pain control/Narcotic dependence*
Methadone (Dolophine, Methadose)
Table 2 Screening/Risk Assessment form (www.sads.org)

Personal History Questions
1. Have you ever fainted or passed out DURING exercise, emotion or startle?
2. Have you ever fainted or passed out AFTER exercise?
3. Have you ever had extreme fatigue associated with exercise (different from other peers?)
4. Have you ever had unusual or extreme shortness of breath during or after exercise?
5. Have you ever had discomfort, pain or pressure in your chest during or after exercise?
6. Have you ever been diagnosed with an unexplained seizure disorder?

Family History Questions
1. Are there any family members who had an unexpected, unexplained death before age 50? (include SIDS, car accident, drowning, other)
2. Are there any family members who died of heart problems before age 50?
3. Are there any family members who have had unexplained fainting or seizures?

Table 3
Resources of additional information on LQTS

Sudden Arrhythmia Death Syndromes (SADS) Foundation
www.sads.org
800-786-7723

Cardiac Arrhythmias Research and Education Foundation
www.longqt.org
800-404-9500

American Heart Association
www.americanheart.org
8-00-242-8721

Heart Rhythm Society
www.HRSonline.org
508-647-0100

National Society of Genetic Counselors
www.nsgc.org
610-872-7608