

Introducing Long QT Syndrome

Long-QT-Syndrome.com

This article was written for individuals seeking simplified information on Long QT syndrome and is introductory in nature. If you are seeking a more detailed description of Long QT syndrome, please visit us at:
www.Long-QT-Syndrome.com

Overview

Long QT syndrome is a rare problem with the heart's electrical system. It is estimated to affect 1 in 5,000 people and result in 3,000 deaths within the United States each year. As the heart pumps blood, its electrical system needs to recharge between beats. In people with Long QT syndrome, the heart's electrical system takes too long to recharge. This makes the heart susceptible to problematic rhythms, often referred to as arrhythmias, which can cause the heart to stop pumping blood throughout the body. If the heart enters into one of these problematic rhythms it may cause a person to become lightheaded or faint. If the heart does not return to its normal rhythm, it may lead to death.

The Underlying Problem

People with Long QT syndrome have a problem with the recharging of their heart's electrical system, but why does it take too long to recharge? In the last fifteen years, scientists and medical doctors have worked hard to answer that question. They have discovered that Long QT syndrome is caused by a defect in a set of very small channels (ion channels) within the heart. These channels allow extremely small, electrically charged particles (ions) to move in and out of the heart's cells. The movement of these electrically charged particles is responsible for both generating and recharging the heart's electrical signals. In people with Long QT syndrome, as the heart recharges, the small channels in the heart's cells don't open and close properly to let the particles move at the correct times. Researchers are still not certain how these channels are connected to the dangerous heart rhythms associated with Long QT syndrome, but it is known that the defective channels make the heart's electrical system unstable as it recharges. When the heart is unstable, it can enter into a problematic heart rhythm.

Problematic heart rhythms come about when the electrical signals in the heart become disorganized and cause the heart to pump blood inefficiently. A few heart rhythms often associated with Long QT syndrome are Torsades de Pointes, ventricular fibrillation, and ventricular tachycardia. If the heart falls into any of these three rhythms death can occur within only a few minutes.

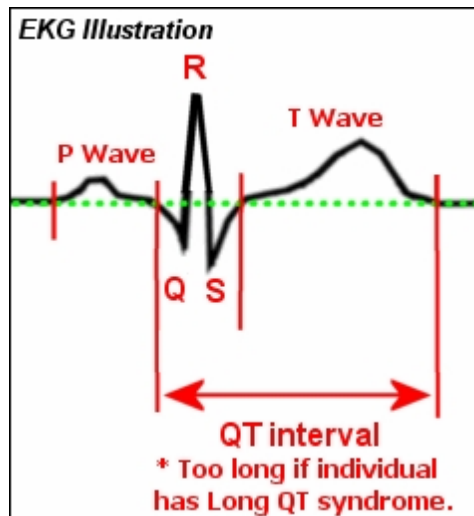
Fortunately, people with Long QT syndrome do not usually experience dangerous heart rhythms on a daily basis. They are, however, in constant danger of experiencing a problematic rhythm. Events that spark the body to release adrenaline are often cited as events that may cause people with Long QT syndrome to experience life-threatening heart rhythms. Therefore, it is usually recommended that people with Long QT syndrome avoid high intensity physical activities and try to remain calm during highly emotional events.

People who have, or are suspected of having, Long QT syndrome often need to avoid certain medications that may cause the heart to enter into problematic rhythms. Several over-the-counter and prescription drugs have been noted for causing potentially deadly heart rhythms in people with Long QT syndrome. It is therefore very important that people with Long QT syndrome avoid taking certain medications. Many of the medications not recommended for individuals who have Long QT syndrome are listed at the Web site:
www.QTDrugs.org

How Long QT Syndrome is Diagnosed

The most widely recognized symptom in people with Long QT syndrome is fainting during or immediately following a highly emotional event or period of intense physical activity. But, this is not to say that a person with Long QT syndrome can not have a problem at anytime. Because people with Long QT syndrome usually appear perfectly normal on the outside and rarely show any symptoms, it can be a very difficult problem to diagnose. In fact, it has been estimated that as high as half of the individuals in the United States who have Long QT syndrome are unaware that they have it.

There are a few key indicators that doctors can use to identify people who may have Long QT syndrome. A family history in which a person's relatives have Long QT syndrome is a strong indicator that a person may have Long QT syndrome, but it is not how the diagnosis is made. There are two important tests that a doctor can do to help determine if a person has Long QT syndrome. One is to give a person a very simple test called an electrocardiogram (EKG). The doctor can use the EKG to determine if an individual's heart is recharging within a normal amount of time by measuring what is called the QT interval. If a person's QT interval is longer than normal, hence the name Long QT, the doctor may diagnose the person as having Long QT syndrome.



A second method of testing for Long QT syndrome is to have a genetic test. To do this, a doctor will remove a small amount of blood from a patient and have it tested to see if they can find any indicators for Long QT syndrome within the patient. Unfortunately, neither test is one-hundred percent foolproof. In general, the EKG is the preferred tool for determining if a person has Long QT syndrome.

Treating Long QT Syndrome

There are currently two major treatment options for people who have been diagnosed with Long QT syndrome; medications (beta-blocker therapy) and ICD/pacemaker implantation. Generally, the second treatment option is reserved for patients at the highest risk of sudden death.

Beta-blockers are the drugs of choice for treating patients with Long QT syndrome. They help block the stimulus that adrenaline applies to the heart. Essentially, beta-blockers prevent the heart's electrical system from spinning out of control during adrenaline releasing events, such as the emotional states associated with the "fright, fight, flight" response. In a 1993 study, it was determined that beta-blocker therapy was effective in reducing Long QT syndrome's symptoms in 76% of 287 affected individuals. Side effects from beta-blockers are generally mild and include low blood pressure, a slowed heart rate, dizziness, tiredness, sudden weight gain, and shortness of breath.

In approximately 25% of Long QT syndrome patients, beta-blocker therapy is not effective in preventing problematic arrhythmias from reoccurring.

Patients who have experienced these so called "breakthrough events" need to be carefully considered for ICD/pacemaker implantations. What is an ICD/pacemaker? It is a mechanical device that cardiologists (heart doctors) can implant underneath a patient's skin. It will monitor a patient's heart rhythm and if a problem occurs, it can administer a powerful shock to their heart (defibrillation). The shock is designed to restart their heart and return it to its proper rhythm... saving the patient's life.

Dealing with An Emergency

If a person with Long QT syndrome experiences a fainting episode the event needs to be treated as an immediate medical emergency. The emergency response system needs to be activated. Advanced life support personnel may be needed to administer CPR or defibrillate the person in order to save the person's life.

Future Outlook

Generally in the case of Long QT syndrome, the patient's care becomes the primary responsibility of the cardiologist, but the family physician should still keep close watch on the patient to ensure that treatments are remaining effective and not causing the patient unnecessary side effects. It is recommended that Long QT syndrome patients see their physician annually. Equipped with the proper knowledge and treatment, patients with Long QT syndrome can expect to live out full and healthy lives.

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