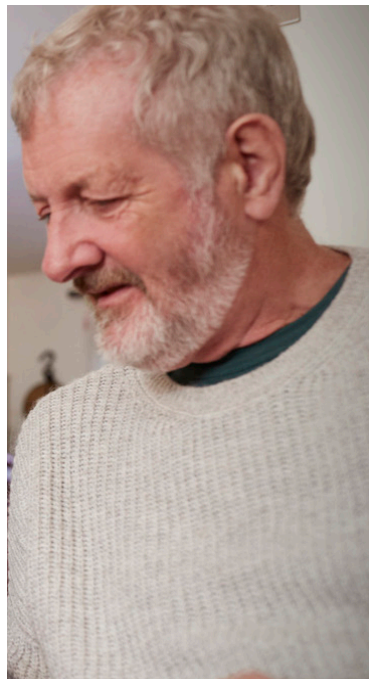




Long QT Syndrome



Working together to improve the diagnosis, treatment
and quality of life for all those affected by arrhythmias

www.heartrhythmalliance.org

Registered Charity No. 1107496

Glossary

Arrhythmia Heart rhythm disorder

Cardiologist A doctor who has specialised in the diagnosis and treatment of patients with a heart condition

Electrocardiogram (ECG) A 12 lead recording of the activity of the heart

Implantable Cardioverter Defibrillator (ICD)
A small device connected to your heart to help treat irregular heart rhythms by using electrical shocks, helping to control life-threatening arrhythmias, especially those that can cause sudden cardiac arrest (SCA)

Pacemaker A small metal device implanted under the skin, which produces electrical impulses to treat an abnormal heart rhythm

Syncope A temporary loss of consciousness and posture, described as 'fainting' or 'passing out'. It is usually related to temporary insufficient blood flow to the brain

Important Information

This booklet is intended for use by people who wish to understand more about Long QT syndrome. The information within this booklet comes from research and previous patients experiences. This booklet offers an explanation of the Long QT condition.

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What is Long QT Syndrome (LQTS)?

Long QT Syndrome (LQTS) is a heart rhythm disorder that can cause a disturbance in the electrical system of the heart. It is often the result of inheriting an abnormal gene which causes an imbalance in molecules that control the electrical impulses of the heart. LQTS can also be caused by medications for other conditions.

Electrically-charged molecules (or ions) such as potassium, sodium and calcium, cross heart cell membranes through specialised ion channels. This generates the electrical activity (depolarization and repolarization) that initiates the heart pumping blood to the rest of the body. The abnormal function of one or more ion channels in LQTS prolongs the repolarization process which predisposes patients to cardiac arrhythmias.

LQTS may result in a very fast abnormal heart rhythm (arrhythmia), known as 'Torsades de Pointes'. When these rapid heartbeats occur no blood is pumped out from the heart and the brain quickly becomes deprived of oxygen, potentially resulting in a loss of consciousness (syncope) and rarely, sudden death.

Arrhythmia in patients with LQTS may be triggered by exercise or stressful situations. Not everyone who has LQTS will have an arrhythmia, but if it does occur it can be fatal. LQTS is an electrical problem so it does not change the shape and function of the heart muscle.

What is the QT interval?

Every time the heart beats, it is due to an electrical impulse that spreads across the heart muscle causing it to contract. The QT interval is measured on the 12 lead surface electrocardiogram (ECG), which is a recording of the electrical activity of the heart.

The QT interval represents the time from the start of the electrical stimulation (depolarization) through the heart's pumping chambers (ventricles), to their resting phase (repolarization). It is a measure of the time from the beginning of the ventricles' contraction until the end of relaxation (see Figure 1). A long QT interval is an abnormally prolonged interval between these two distinct points on the ECG.

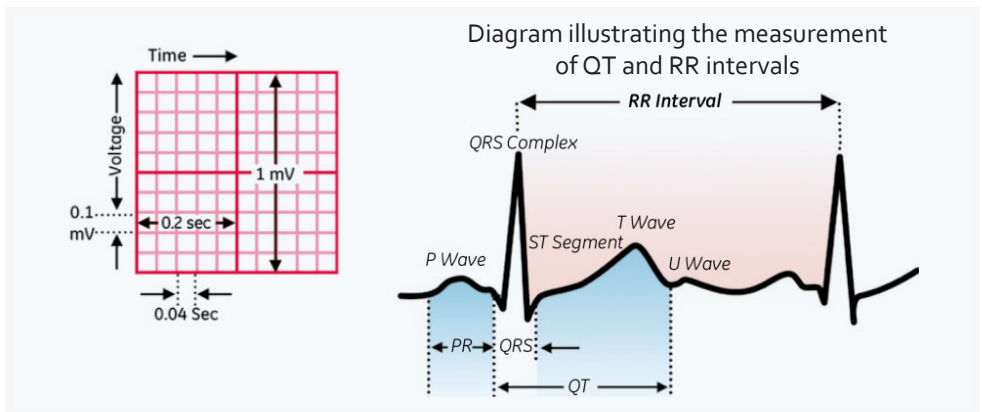


Figure 1

What are the symptoms?

It is estimated that up to 50% of people with LQTS do not have symptoms. They may be aware of their condition only from results of an ECG performed for an unrelated reason, because they have a family history of LQTS or because of genetic testing results.

People with LQTS may start to experience symptoms in childhood, although this is not always the case. Symptoms include:

- Sudden, unexplained fainting, particularly when in response to a stressful situation. This can often be misdiagnosed as having a 'hysterical reaction'.
- Unexplained seizures. A sudden loss of consciousness may be mistaken or misdiagnosed as an epileptic seizure.
- Sudden cardiac arrest or death in the absence of any structural heart disease or other cardiac problems. Approximately 1 in 10 sudden cardiac arrests or death are the first sign of LQTS.

What causes Long QT Syndrome?

LQTS can be inherited or acquired. Acquired LQTS is usually due to the administration of certain medications. Many groups of common medications can cause a prolonged QT interval including certain antibiotics, antihistamines, antidepressants, antipsychotics and heart medications. A comprehensive list of medications can be obtained from your physician. Inherited LQTS is caused by mutations of certain genes and can be passed onto family members. The exact frequency of inherited LQTS is not known; however, LQTS overall is estimated to affect approximately 1 in 2,000 people. There are several different types of inherited LQTS and your cardiologist may be able to tell you which type you have. The three most common types of inherited LQTS are called LQTS 1, LQTS 2 and LQTS 3.

There are 17 different subtypes of LQTS associated with at least 15 genes associated with LQTS discovered so far, and hundreds of mutations within these genes have been identified. Mutations in three of these genes (LQTS 1, 2 and 3) account for about 70 to 75 percent of LQTS cases. The type of LQTS may be identified by genetic testing. In types 1 and 2, the potassium channels within the heart cause the problem. In these types, arrhythmia may be triggered by exercise or by emotional stress. In type 3 it is the sodium channel that is affected and a low heart rate during sleep or rest may be the trigger for arrhythmia.

What are the risks?

LQTS is a rare condition, experts would suggest that approximately 1 in 2,000 people are affected but this is not certain as it may never be diagnosed. You may be at risk if anyone in your family has had unexplained fainting episodes or seizures, or an unexplained sudden cardiac death. You are at increased risk if you have a first-degree relative with known LQTS.

You are also at risk if you are taking any medications that prolong the QT interval. Your doctor can inform you whether any of your medications can do this. Certain metabolic conditions can also cause prolonged QT interval. People with low potassium, magnesium or calcium blood levels; such as those with the eating disorder anorexia nervosa, or have had diarrhoea or vomiting, are also at increased risk of prolonged QT interval.



Test and diagnosis

ECG

An ECG is a simple tracing of the heart's electrical activity. It involves attaching electrodes to the chest and limbs and a recording is made. It may reveal a long QT interval which may suggest that it is more likely that you have LQTS. Not all people with LQTS have a prolonged QT interval on their resting ECG and it may be necessary to undertake several ECGs over a period of time, or have a period of continuous monitoring using a portable heart monitor.

Exercise tolerance test

Some people may only have a prolonged QT interval when exercising so it may be necessary to have ECG monitoring done while exercising on a treadmill. This is normally the best way for your doctor to find out if you have LQTS. It can also help your doctor find out which type of LQTS you might have.

A non-exercise (medication) stress test

An ECG test is performed while you are given a medication that stimulates your heart in a similar way to exercise. The medication is given through a vein in your arm and may include epinephrine (adrenaline). Doctors monitor the effects of the adrenaline on the way your heart recharges. This test can unmask what is known as concealed or borderline LQTS and mimics the heart's response to a sudden burst in adrenaline.

Neurological test

An electroencephalogram (EEG) test looks for neurological causes of fainting, such as a seizure disorder. The procedure measures the waves of electrical activity the brain produces. Small electrodes attached to your head pick up the electrical impulses from your brain and send them to the EEG machine, which records brain waves. An EEG looks for other conditions that LQTS may be misdiagnosed as, e.g. epilepsy.

Medical history and assessment

Your cardiologist will ask you a series of questions regarding your symptoms including faints, blackouts and palpitations and about your family history. Please mention anyone who has died unexpectedly and suddenly. It can be helpful if you start talking to your family and asking about their medical health, particularly if they have any symptoms which might be related to LQTS. Your cardiologist will also want to know what medications you are on. You will be asked about how much exercise you do. You will also be asked if you have any conditions that may cause the potassium levels within your blood to fall, which can trigger arrhythmia. These conditions could include excessive vomiting or diarrhoea, anorexia nervosa or certain thyroid problems. You may have blood taken to check your blood potassium levels.

Genetic testing

If your cardiologist thinks you may have LQTS or knows you have LQTS based on your clinical tests (e.g. ECG, exercise test and medical history), they may refer you to a genetic specialist. The genetic specialist can perform a genetic test on you to try and identify if you have a genetic alteration which is causing your LQTS. However, these tests do



not always reveal a genetic alteration and therefore, a negative test does not necessarily mean that you do not have the condition. This is partly because not all the genetic alterations, that cause LQTS have been identified yet. If your test identifies a genetic alteration, your relatives can be tested for the same genetic alteration. The genetic test should be used in conjunction with an ECG and exercise test. The presence of a genetic alteration does not tell us whether you will have symptoms from long QT. The ECG, exercise test and medical history normally provides your doctor with more information in trying to determine whether you will develop symptoms.

Treatment

The main aim in treatment is to prevent loss of consciousness and a life threatening arrhythmia from occurring. There is no cure for LQTS but treatment options include: medications, medical devices, surgery or lifestyle changes. Treatment will be dictated by what type of long QT you have and what is most suitable for you.

It is wise to inform other people if you have LQTS so that they know to call for urgent medical help if you were to faint. Identity bracelets are available from certain charities which carry medical information about you. Your local arrhythmia nurse or your cardiologist may be able to give you more information about this. Alternatively, ask Arrhythmia Alliance for further details.

There are many medications which might affect the heart rhythm in patients with LQTS. These include some over-the-counter cough or cold remedies (decongestants) and some antibiotics. Other drugs that might affect the QT interval include some antidepressants, some treatments for fungal infections, and drugs for heart rhythm disorders. If you are prescribed any medicines, always check with your doctor and pharmacist that it is safe for a patient with LQTS to take these medicines. Some herbal remedies are also to be avoided (e.g. St John's Wort). Please take care with herbal remedies and ask your doctor for specific advice.

A list of drugs currently known to affect Long QT are available now on www.crediblemeds.org. This list will not be exhaustive as newer drugs are becoming more available. Always inform anyone who is prescribing you medication that you have LQTS as there may be newer drugs on the market which may have not have been added to the website.

Medications

Beta blockers: These drugs slow the heart rate and make the dangerous rhythm associated with LQTS less likely. They work by blunting the way a LQTS-affected heart reacts to adrenaline in times of stress, fear or exertion.

Mexiletine: In people with a form of LQTS called LQT₃, taking this antiarrhythmic drug in combination with a beta blocker may help shorten the QT interval.

Potassium: Potassium is a mineral in your body that is important for your heart's electrical system. Potassium supplements may improve the heart's recharging system and may be helpful for people with certain forms of LQTS.

Medical devices and surgery

A pacemaker or implantable cardioverter defibrillator (ICD): These devices are implanted under the skin of your chest and stop a potentially fatal arrhythmia. A pacemaker produces electrical impulses to treat an abnormal heart rhythm.

An ICD continuously monitors your heartbeat and will deliver electrical shocks to restore a normal heart rhythm when necessary.

Left cardiac sympathetic denervation surgery: Specific nerves in your chest controlling your heart rhythm are surgically removed which significantly reduces the risk of sudden death. This surgery is generally reserved for people considered at high risk of sudden death, people who do not tolerate the medications or have a fainting spell despite their medications.

Lifestyle changes

If exercising triggers your fainting, you may wish to avoid strenuous activity.

Depending on the type of LQTS, you may be advised not to take part in certain types of sporting activities. It may be that competitive sports are not advisable. However, this will need to be discussed with your doctor or specialist nurse, on an individual basis. Furthermore, your career options will need to be discussed, as there may be certain restrictions within sports, the police force or the armed forces.

You may also wish to have a diet containing foods rich in potassium, and inform your doctor of any illness which could cause your potassium levels to fall (as mentioned earlier).

Additionally, you may be advised to avoid using alarm clocks, lowering doorbell volumes, and turning your telephone off at night.

It is wise to inform other people if you have LQTS so that they know to call for urgent medical help if you were to faint.

Sexual intercourse does not appear to increase the risk of LQTS. Neither pregnancy nor delivery are associated with increased risk of symptoms in women with LQTS. However, there can be an increased risk of arrhythmia after the birth. This increased risk can last for up to 9 months after the delivery. Nonetheless, your doctor will want to monitor you closely both during your pregnancy and after if you have inherited LQTS.

WARNING Recreational drugs such as Ecstasy and Cocaine are particularly dangerous in patients with LQTS, and CAN BE FATAL. Patients with even mild LQTS should NEVER experiment with these drugs.

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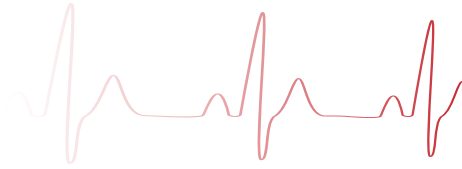
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This booklet has been written to support those diagnosed with an arrhythmia and their carers, who struggle to find information on this condition.

Without donations and fundraising, we would not be able to provide support through our award-winning resources and helpline.

Please donate to support our vital work at
www.heartrhythmalliance.org/aa/uk/get-involved/donate



Working together to
improve the diagnosis,
treatment and quality
of life for all those
affected by arrhythmias



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**"After the initial shock of being told I
have LQTS, I was able to make a few
small changes to my lifestyle with the
help of this booklet"**

Michelle, Warwickshire

To view our patient resources, scan the
QR code below:



Please remember that this publication
provides general guidelines only. Individuals
should always discuss their condition with
a healthcare professional. If you would like
further information or would like to provide
feedback, please contact Arrhythmia Alliance.

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If you would like further information or would like to provide feedback please contact Arrhythmia Alliance.