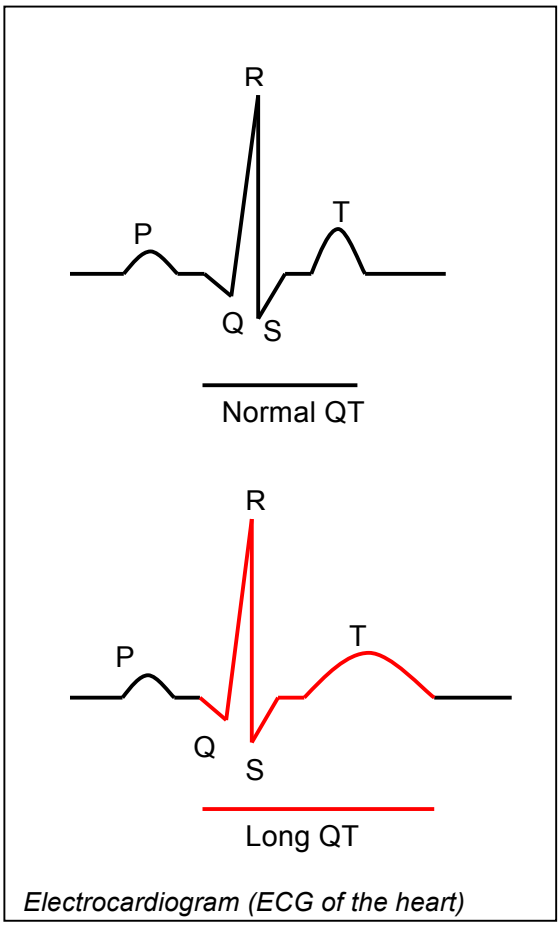
# Long QT Syndrome (LQTS)

Long QT syndrome (LQTS) is a condition in which there is abnormal electrical activity in the heart. It is an uncommon condition that affects the heart of otherwise fit and healthy people, causing fainting or collapse, sometimes resulting in death. It most commonly occurs in children and young adults, typically in their teens or 20s. When an affected person has affected relatives, the condition is said to be 'genetic', 'familial' or 'inherited'.

Treatment is available once a diagnosis has been made, However, diagnosis can be difficult, because not all people affected by LQTS have symptoms and the testing can sometimes be normal.

Usually, LQTS can be seen on an electrocardiogram (ECG). Every heartbeat is triggered by an electrical signal that tells the heart's muscle cells to contract. After contracting, these cells must recover – or relax – before the next heartbeat starts. The amount of time taken by these cells to recover can be measured on an ECG and this is called the QT interval. During the last part of this interval, the heart is vulnerable and electrically unstable. People with LQTS have an abnormally long QT interval and if the next electrical signal arrives before the muscle cells have completed their recovery period, a dangerously fast heart rate can occur, leading to a fall in blood pressure and loss of consciousness.



*Electrocardiogram (ECG of the heart)*

People with LQTS are sometimes identified after an unexplained fainting episode. These episodes are usually associated with surges of adrenaline, such as a sudden loud noise, intense emotional reactions, awakening from sleep or during intense physical activity, especially swimming. As some medications can further prolong the QT interval or increase the risk of an abnormal fast heart rhythm, patients with LQTS must avoid certain medications. All patients with suspected or confirmed LQTS should be familiar with the full list of drugs that must be avoided. This list is available at: http://www.qtdrugs.org

**How it is inherited**

LQTS is inherited as an autosomal dominant disorder (see section on Genetic Inheritance for more information).

**Genetic testing**

Genetic testing is now commercially available and involves screening six genes that can cause the condition. The gene alteration will be identified in approximately 70-80% of families.

**Treatment**

Treatment varies according to severity. Most people who carry a gene for the LQTS will not be severely affected and may never have symptoms. However, all gene carriers

should avoid certain drugs and medications that could trigger a heart rhythm disturbance (www.qtdrugs.org). Many will be advised to make lifestyle changes e.g. avoid swimming or competitive sport (see information sheet on Lifestyle Modifications). Lifestyle modifications should always be discussed with your cardiologist. Taking beta-blocker medication every day reduces risk significantly, especially in boys. Those few people with LQTS at highest risk of cardiac arrest may be advised to have a pacemaker-defibrillator (known as ICD or implantable cardioverter defibrillator). A small operation to cut nerves to the heart can also be helpful (left cervical sympathectomy).

Everyone with LQTS should see their cardiologist at least once a year.

**For more information**

To find out more about genetic heart conditions and the latest research news, please visit the Australian Genetic Heart Disease Registry at www.heartregistry.org.au or the Cardiac Inherited Diseases Group (New Zealand) at www.cidg.org