

## Long QT syndrome

Information for individuals and families affected by long QT syndrome.

### What is long QT syndrome?

Long QT syndrome refers to a condition where a person has a particular abnormality of the heart's electrical system, without any other heart abnormality. The name refers to the way the condition alters the picture of the electrocardiogram (ECG or electrical tracing of the heart). The ECG shows a longer than usual interval between two points on the tracing. These points are known as 'Q' (first arrow) and 'T' (second arrow).



Normally, a tiny electrical current triggers the muscular contraction of the heart, allowing it to pump blood. This current passes from the top to the bottom of the heart. As this electrical 'wave' passes through the walls of the heart, the muscle fibres in the walls begin to contract. After this has happened, the individual heart muscle cells spontaneously return to their normal resting electrical state to wait for the next heartbeat. In long QT syndrome, this 'return to rest' takes a longer period of time than for a normal heart.

There are various causes of a long QT interval on an ECG, such as certain drugs that can interfere temporarily with the electrical system of the heart. In most cases it is due to an abnormal gene that may be shared or inherited by family members. For this reason, long QT syndrome is called a *genetic* condition. Rarely, it may be associated with deafness in babies.

The hearts of people with long QT syndrome work normally most, or all, of the time. There is, however, a greater risk than normal that the heart will spontaneously get out of rhythm and stop pumping regularly. In some people this leads to fainting spells or blackouts. In some cases, it can lead to cardiac arrest and sudden death. However, the risk of sudden death can be considerably reduced with simple treatments.

### How is the condition inherited within the family?

Long QT syndrome is a genetic condition that is usually inherited in a 'dominant' manner. This means that each child of an affected parent has a one in two (50 per cent) chance of inheriting the syndrome. If a child has not inherited the faulty gene causing long QT syndrome, he or she cannot pass it down to his or her children. In this sense, it does not 'skip' a generation.

In rare cases, it is possible to inherit the gene fault that causes the syndrome, but for some reason the gene fault does not always produce an obvious

abnormality on every ECG recorded. This happens in many other genetic conditions; it is not peculiar to long QT syndrome. However, this makes it very difficult to make a diagnosis in some people, or to know if they are at risk of the problems associated with the syndrome. It also makes it difficult to work out the risk of inheritance in their children. This is why it is always important for health professionals to take a detailed family history, as this full picture can sometimes help with making a clear diagnosis. Once the specific genetic defect has been found in a family this difficulty can be resolved, but at the moment scientists are only aware of about 60 per cent of the genes that cause long QT syndrome.

Once a new diagnosis of long QT syndrome has been made, or if there is a suspected diagnosis, it is recommended that a cardiologist or a paediatric cardiologist with expertise in the condition checks all children, siblings and parents of the person with the syndrome. An ECG will be required to see if the condition has been inherited and other tests, such as an exercise test, may be necessary.

Sometimes it is very difficult to make a firm diagnosis of long QT syndrome on the basis of the ECG recording because some people have a 'borderline' lengthening of the QT interval. While this may be normal for some, it may indicate the syndrome in others. At present, experts recommend that people with a borderline QT interval, who are part of a family with definite long QT syndrome, be treated in the same way as their relatives who have a definite diagnosis.

### **What are the risks and outcomes of long QT syndrome?**

Most people with long QT syndrome survive and live normally. This has enabled the condition to be transmitted from generation to generation through affected families.

Some people with the syndrome have recurrent blackouts. Unfortunately, these are sometimes misdiagnosed as common fainting or dizzy spells, or epilepsy. These blackouts sometimes, but not always, occur under conditions of exertion, emotion or excitement. Episodes while swimming are common. Less commonly, these can occur during sleep. People who experience blackouts are at risk of sudden death. Fortunately, this risk is considerably reduced with simple modern treatments.

### **Can one predict who is at highest risk of serious problems?**

People who are at highest risk of serious problems associated with long QT syndrome are those who have recurrent blackouts, those with a very long QT interval, and females more than males. Unfortunately, while these indicators can be used to identify most people who may experience serious problems, there is still a small risk even for those people who do not have these indicators.

### **What can be done about the problems of long QT syndrome?**

Beta-blockers, which are medications widely used to treat high blood pressure and other conditions, are also used in long QT syndrome. This is because they are known to reduce or eliminate blackouts and reduce the chances of sudden death in long QT syndrome.

### **What are the effects of beta-blockers?**

Beta-blockers work by dampening the bursts of nerve impulses that go to the heart and excite its electrical system during exercise, or emotion. They slow the heart rate and reduce blood pressure and have been used for many years with no serious long-term effects. In large doses, beta-blockers may cause faintness (especially on standing up suddenly), lethargy and exercise intolerance. Usually these symptoms improve if the dose is reduced slightly. The dose should be just a little below that which causes these side effects.

Beta-blockers may make asthma worse and this can be a difficult problem for those with long QT syndrome. Certain types of beta-blockers ('selective beta-blockers') are generally

suitable for asthmatics. However, if asthma is already severe, or becomes severe on beta-blockers, it is probably better not to take them unless the person is judged to be at high risk of the serious heart symptoms described.

It is very important that people with long QT syndrome take these drugs if possible, but it is equally important to establish a dose that does not cause major side effects. Medications and their dosages should only be changed in consultation with the doctor who is treating the condition.

As beta-blockers slow the heart rate, those with already slow heart rates may need a permanent artificial pacemaker to allow treatment with these drugs. For information on artificial heart pacemakers, contact Heartline on 1300 36 27 87 (local call cost) or visit [www.heartfoundation.com.au](http://www.heartfoundation.com.au) .

For people who continue to have symptoms despite treatment, and for those who are unable to take beta-blockers, an operation called 'stellate ganglionectomy' may be carried out. This operation decreases the nerve supply to the heart and is usually done only in symptomatic individuals.

### **Cardiac arrest and CPR**

Even with treatment, people with long QT syndrome may be at greater risk of sudden death than other individuals.

If cardiac arrest occurs (the person becomes unconscious and has no pulse), cardio-pulmonary resuscitation (CPR) must be given until help arrives. As a precaution, it is recommended that everyone in long QT families knows the principles of CPR.

If a person develops a life-threatening arrhythmia and suffers cardiac arrest, a piece of equipment called a defibrillator can be used to deliver a controlled electric shock or series of shocks to the heart and restore a normal rhythm. Defibrillators are carried in specially equipped ambulances and are also located in hospitals and some clinics. Increasingly, a special type of 'automatic' defibrillator is becoming available in other locations including some commercial aircraft and where large numbers of people congregate, such as football grounds. These machines are designed for use by non-health professionals, and may also be considered for the home, particularly for families with a number of individuals affected by long QT syndrome.

People who are at very high risk may receive an implantable defibrillator, which is placed in the chest and automatically delivers an electrical current to enable the heart to revert to normal should it develop a dangerous abnormal rhythm. For more information on implantable cardiac defibrillators, contact Heartline on 1300 36 27 87 (local call cost) or visit [www.heartfoundation.com.au](http://www.heartfoundation.com.au) .

### **Are there any activities or drugs that should be avoided?**

#### ***Activities***

For those with the condition, vigorous or physically demanding sports may need to be avoided.

#### ***Medications***

Some drugs are known to prolong the QT interval in any person who takes them. People with long QT syndrome should avoid these types of drugs which include:

- Cardiac – Many anti-arrhythmic drugs, such as Quinidine, Sotalol, Amiodarone
- Psychiatric – Phenothiazine drugs, such as Thioridazine (Melleril) and some anti-depressants, e.g. Amitriptyline (Tryptanol)
- Antibiotics – Erythromycin, Ketoconazole and Pentamidine
- Antihistamines – Astemizole (Hismanal), Terfenadine (Teldane)

- Diuretics (fluid tablets) – These may cause a low level of potassium in the blood, which prolongs the QT interval
- Gastrointestinal – Cisapride (Prepulsid), Ondansetron.

All people with suspected or confirmed long QT syndrome should be familiar with the full list of drugs that must be avoided. If unsure, people with long QT should check with a doctor or pharmacist before taking any new medication. Information can also be obtained by contacting the Medicines Line on 1300 88 87 63 (local call cost).

### ***Other issues***

Liquid protein diets, Chinese herbs and exposure to organophosphates could aggravate long QT syndrome.

Alarm clocks are better set to a gentle music station rather than a startling alarm.

If a person with long QT syndrome has diarrhoea, their blood levels of potassium may fall and they may require additional potassium and magnesium supplements.

### **What does the future hold?**

Worldwide genetic research into affected families has now identified a number of different genes which cause long QT syndrome. This vital research is leading to significant advances to help identify those affected and to provide treatment options that aim to treat the cause of the condition, rather than its effects.

Identifying the genetic defects in a particular family will finally assist doctors to make a more accurate diagnosis in other relatives who are at risk of inheriting the condition. Researchers are also beginning to understand what causes the heart condition, through the identification of particular genes. With such knowledge, eventually it may be possible to 'silence' or even repair the abnormal gene. In the future, genetic typing of each family may be used to select a specific, more effective treatment for that family.

With this research, and a better understanding of the way abnormal genes change the electrical properties of heart cells, it is hoped that before too long more specific and effective treatments will replace beta-blockers. These treatments will help prevent the detrimental effects of long QT syndrome.

### **Further information**

The following organisation can provide further information and support for those affected by long QT syndrome.

Australian Sudden Arrhythmia Death Syndromes  
SADS Foundation  
PO Box 19  
Noble Park Victoria 3174  
Australia  
Website: [www.sads.org.au](http://www.sads.org.au)  
Email: [info@sads.org.au](mailto:info@sads.org.au)

For information on a range of other heart health topics, contact Heartline, the Heart Foundation's national telephone information service, on 1300 36 27 87 (local call cost) or visit [www.heartfoundation.com.au](http://www.heartfoundation.com.au).

© National Heart Foundation of Australia. February 2004.

The Heart Foundation would like to thank Dr Andrew Davis, Paediatric Electrophysiologist, The Royal Children's Hospital, Victoria, Dr Brian Bailey, Royal Prince Alfred Hospital, Sydney, and Dr Jitu Vohra, Cardiologist, The Royal Melbourne Hospital, Victoria, for their assistance in preparing this information.

**This information sheet has been developed for educational purposes only. The information should not be used for diagnosing or treating a health problem or a disease and should not be substituted for professional care. Please consult your health care provider if you have, or suspect you have, a health problem.**