FACTSHEET

Long QT syndrome

What is Long QT Syndrome?

Long QT syndrome (LQTS) is a heart rhythm disorder, which can cause periods of abnormally fast heart rhythm called arrhythmias. Long QT Syndrome is typically present at birth, but it is often only detected in children and young adults. In most cases Long QT Syndrome can be detected using a test that measures the heart's electrical activity, called an electrocardiogram (ECG).

The pumping action of the heart is a result of a complex electrical system that triggers the heart muscle to contract in the right order and at the right time. After squeezing, the heart muscle needs to relax before the next heart beat begins. The QT interval is the amount of time needed by the heart muscle cells to recover. An ECG can measure the length of the QT interval. In people with Long QT Syndrome, as the name suggests, this interval is longer than normal.

During the heart muscle cell recovery time, the heart is electrically vulnerable and if another electrical impulse arrives during this time it can result in a dangerously fast rhythm. In many forms of Long QT Syndrome, this is more likely to occur if the person is doing strenuous activity. If this fast heart rhythm occurs, little blood is pumped from the heart which can lead to a fall in oxygen supply to the body and brain and a loss of consciousness (fainting episode). If this episode is not recognised and treated immediately it may be life-threatening.

Some children are born with Long QT Syndrome because they have inherited a gene change associated with Long QT Syndrome from one or both of their parents. Parents may have been unaware they had the condition because they may not have had any symptoms.

Long QT Syndrome can also result from a gene change that has happened after conception. If this is the case, the gene change can be passed on to the affected child's own children in the future, but their siblings are usually unaffected.

How is it treated?

The treatment for Long QT Syndrome is aimed at reducing the risk of the arrhythmia from occurring. Lifestyle changes and medication are very effective and will be tailored to the individual.

Medications:

The first treatment is usually a beta-blocker medication such as propranolol, atenolol or nadolol. This group of medications helps control irregular heartbeats and can slow the heart rate.

Lifestyle modifications:

Your Cardiologist will provide information specific to your child regarding what lifestyle changes or

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modifications you may need to adhere to. For most, this will mean avoiding strenuous activity, especially competitive sports and swimming, and for some they will need to avoid loud noises that may startle them or awake them when sleeping.

Implantable cardioverter defibrillator:
If beta-blockers are not effective in managing your child's heart rhythm, your child may need to have an operation to implant an implantable cardioverter defibrillator (ICD). This device is similar to a pacemaker but is also able to deliver an electrical shock if it senses that the heart is beating in a potentially dangerous rhythm. This can help return the heart to normal rhythm and get it pumping again.

Automatic External Defibrillator:
An Automated External Defibrillator (AED) is a portable device that is used to deliver an electric shock in order to make the heart rhythm normal again. In this way it is similar to an ICD, however the AED is an external machine that has adhesive pads that need to be applied to the chest in order for the shock to be delivered. The current devices are all preprogrammed to analyse the rhythm once the pads are applied and will audibly instruct the user what action is required. It is best to speak to your Cardiologist about whether this device is recommended for your child.

Medications to avoid

It is important to note that there are many medications that may further prolong the QT interval or increase the risk of the abnormal fast heart rhythm; therefore there are many drugs that need to be avoided in people with Long QT Syndrome. A regularly updated list of such drugs can be found at www.crediblemeds.org.

Helping detect Long QT Syndrome in other family members

Once a diagnosis of Long QT Syndrome has been made in your family, it is very important that other direct family members are checked for the possibility that they may also have the condition. Your cardiologist or geneticist will advise on how other family members can be tested.

In the first instance, this may mean organising an ECG for other family members. However, an ECG is not always 100% accurate. If a gene fault in the family has been identified, then other family members can be tested to find out if they carry the same gene change. The Inherited Arrhythmia Clinic can help with the coordination of these tests.

It is important to know that Long QT Syndrome in the same family can affect family members very differently. Some people with Long QT Syndrome will be completely "asymptomatic". This means that they may never have any symptoms in their lifetime. Others, even within the same family, can be more severely affected.

How we can help

The Heart Centre for Children is dedicated to supporting families with children diagnosed with Long QT Syndrome. The Inherited Arrhythmia Clinic (IAC) at the Heart Centre for Children, The Children's Hospital at Westmead, offers a comprehensive range of services for families with this diagnosis and other arrhythmia. Please refer to the IAC factsheet at www.heartcentreforchildren.com.au

Contacts:

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