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Inherited long QT syndrome (LQTS) affecting the heart

Cardiology and Clinical Genetics

Information for Patients

Produced: July 2021
Review: July 2024
Leaflet number: 405 Version: 2

What is long QT syndrome?

Long QT syndrome (LQTS) is a condition that causes problems with the electrical activity of your heart. This is usually due to inheriting a faulty gene from your mother or your father. This results in the chemicals such as sodium and potassium, becoming out of balance. As a result, it takes longer for your heart to reset itself electrically after every beat.

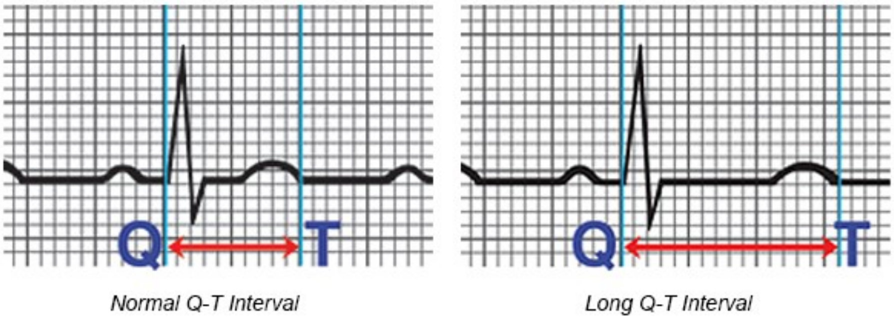
This can show up on a test called an electrocardiogram (ECG) by a measurement called the **QT** interval which is longer than normal, called a 'long' QT interval (see picture below). If this occurs you can be at higher risk of having abnormal and sometimes life threatening heart rhythms that start in the bottom chambers of your heart (ventricles).

How is long QT syndrome diagnosed?

Your doctor may have found out that you have LQTS when investigating you for symptoms such as blackouts or fainting, dizziness or heart palpitations, or from a routine ECG.

Sometimes, a diagnosis is made after screening an individual who has a known family history of LQTS or a family history of unexplained sudden cardiac death.

Some people may have no symptoms, but are found to have LQTS from genetic testing, when a specific genetic change associated with LQTS is known in their family.



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What tests will I need?

Tests may include an ECG, 24 to 48 hour home ECG monitoring, and an exercise ECG test, all of which are painless and involve attaching sticky labels (sensors) to your chest.

Often genetic testing is needed which can be done on a small amount of your blood or sometimes a small amount of your spit. You may have inherited a gene that causes long QT, but your QT interval is within normal range. This doesn't mean that you don't have the condition, it's just not seen from other tests.

You will usually be offered periodic review in the cardiology clinic for monitoring your symptoms and an ECG, and to assess response to treatment.

What do my genetic test results mean?

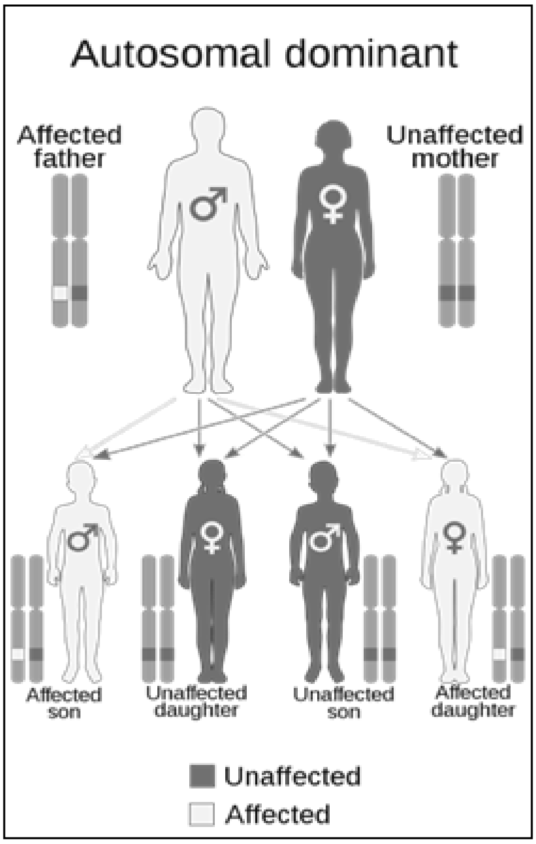
Our genes are like 'instructions' that control how we grow and all the functions within our body. Most genes come in pairs, where one copy of each pair is from our mother and the other is from our father. A change (alteration) in a certain gene may result in a specific medical problem that may be inherited within a family.

The genes KCNQ1, KCNH2 and SCN5A are important for electrical function of your heart. An alteration in one copy of these genes may lead to someone developing an inherited type of LQTS (LQTS type 1, 2 or 3 respectively).

Why is my genetic diagnosis important for my family?

In a family with LQTS linked to a known genetic alteration, there is a 1 in 2 (50%) chance that a child may inherit the altered gene from an affected parent. This is known as autosomal dominant inheritance, as shown in this picture.

Both men and women can inherit LQTS. Each person may be affected by LQTS differently, even within the same family. The effects may vary depending on how severe the LQTS is and the age when any symptoms develop.



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If you have a family history of LQTS or there is a known associated genetic alteration in your family, or there is an unexplained sudden death particularly at a young age (under 40 years), please talk about this as a family and ask your GP for a referral to Cardiology and/ or Clinical Genetics for an assessment.

Can long QT syndrome be treated?

At the moment there is no cure for LQTS. Your heart doctor (cardiologist) will talk to you about the best way of looking after your condition.

Most people with LQTS will need treatment with medicines called beta blockers to reduce the risk of blackouts or your heart stopping (cardiac arrest). Sometimes other drugs are used.

If you are at high risk, especially if you've had a cardiac arrest you may need to have an implantable cardioverter defibrillator (ICD) fitted.

In some cases of long QT syndrome, surgery may be needed on the left side of your neck to treat the nerves and control the flow of chemicals into the heart. This is known as a sympathectomy.

Living with long QT syndrome

With the right treatment, it should be possible to lead a fairly normal quality of life and have a normal life expectancy. However, you may need to make some lifestyle changes to lower your chance of having further problems.

We advise everyone with LQTS to consider the following advice:

- Avoid energetic exercise or playing competitive sports, particularly swimming if you have LQTS type 1.
- Avoid startling noises, such as alarm clocks if diagnosed with LQTS type 2.
- Avoid stressful or very upsetting situations if diagnosed with LQTS type 3.
- Make sure you keep well hydrated (e.g. if you have an illness which causes diarrhoea).
- Consider adding more potassium-rich foods to your diet or having prescribed potassium supplements. Good food sources of potassium include bananas, certain meat and vegetables, nuts and seeds (further advice can be found at www.nhs.uk).
- Always tell medical staff that you have long QT syndrome, especially if you are given any medication (prescribed or over-the-counter), as some medication can increase the QT interval and increase your risk of developing abnormal heart rhythms. The CredibleMeds® website https://www.crediblemeds.org/ (free, but registration required) has a complete and updated list of medications to avoid.
- Consider seeing a genetic counsellor and counselling before planning a pregnancy, given the inherited nature of this condition.
- Let your family members know that they would benefit from heart (cardiac) screening for LQTS.

Where can I find more information or support?

- Sudden Arrhythmia Death Syndromes Foundation: https://www.sads.org/
- British Heart Foundation: https://www.bhf.org.uk/
- Cardiac Risk in the Young (CRY): www.c-r-y.org.uk

Contact details

Cardiology: 0116 258 3297

Genetics: 0116 258 5697

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