

What is Long QT Syndrome?

The Long QT Syndrome (LQTS) is an abnormality of the heart's electrical system. Usually the heart itself is structurally normal and squeezes normally. The primary problem is in the electrical characteristics of the heart muscle cells. To be more specific, the sodium and potassium ion channels of the cell membrane do not work normally. It can only be diagnosed with an EKG.

LQTc is a measurement made on a regular electrocardiogram (EKG) and looks at the amount of time it takes for the heart muscle to "recharge". We measure it from the beginning of the "0" wave to the end of the "T" wave. It takes into account the heart rate. One can think of heart muscle cells as tiny rechargeable batteries that have a certain amount of time to recharge for the next heart beat. In people with LQTS the recharge time is too long and this can create the possibility of developing potentially dangerous irregular, very rapid heartbeats. These "bad" heartbeats can cause dizziness, fainting and sometimes sudden death. The name of this bad heartbeat is Torsade de Pointe.

LQTS was first described in 1957 and is frequently inherited and runs in families. Occasionally it will occur sporadically. There are two types of familial LQTS. The most common is the autosomal dominant Romano-Ward type. The other type is Jervell-Lange-Nielsen which is autosomal recessive. This type of LQTS is associated with hearing loss. Recently there has been a lot of exciting work on the genetics of LQTS. Thus far there has been 4 genes associated with it. KVLQT1 on chromosome 11, HERG on chromosome 7, SCN5A on chromosome 3 and mm K on chromosome 21. The frequency of LQTS is felt to be about 1 in 5,000 in the United States and probably accounts for 3,000 to 4,000 deaths per year. On occasion LQTS can sometimes be caused by certain medications. Sometimes newborn babies that have not been fed much will show it temporarily on an EKG. Luckily, most of these babies' EKGs will become normal in a few weeks. This is because the calcium and magnesium levels normalize after they have been fed.

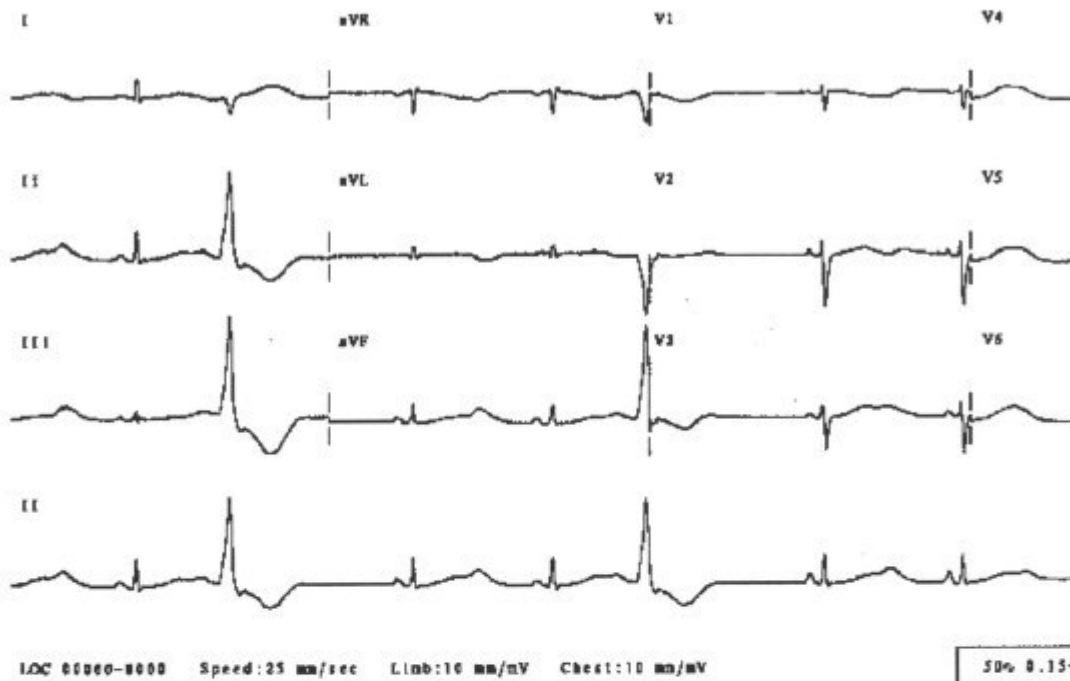
The most common symptom is fainting or syncope, typically during physical activity like exercising or being emotionally upset. Loud noises can trigger it. The symptoms usually start around the teenage years, but in some cases this can be diagnosed in babies (seen with slow heart rates or with heart block). Sometimes the first symptom is sudden death. The sudden loss of consciousness during or shortly after exercise is very suspicious for LQTS. A history of family members fainting suddenly or dropping dead especially if young should raise the suspicion for LQTS. The reason a person faints is because the heart starts to beat so fast that it cannot pump enough blood to the brain, if this continues long enough it will be lethal.

The treatment for LQTS is usually effective in preventing sudden death for most patients. **There is no cure.** The most common medication we use is called beta-blockers (Tenormin, Inderal, Corgard). We also use another medication called Mexiletine as a secondary drug. Many patients will need to have a pacemaker placed and some patients require the placement of an internal defibrillator. This device can help to terminate lethal irregular heartbeats. Treatment choices depend on how long the LQTS is, the symptoms of the patient, and the family history. The upper limits of normal for the LQTS is 0.44 seconds. LQTS greater than 0.60 seconds is extremely dangerous even with treatment. Almost anyone with a LQTS of more than 0.45 seconds should be treated. All family members of a patient with LQTS should have an EKG done.

Drugs to avoid if you have include epinephrine, Erythromycin, Benadryl, Bactrim, Septra, Quinidine, Propulsid, Nizoral, Diflucan, Elavil, Tofranil, Haldol, Risperdal, Betapace. There are others.

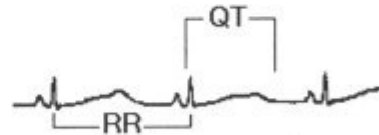
If you have any questions, please ask one of the doctors.

A lady with Romano-Ward syndrome.



Long QT interval

- The QT interval normally varies with heart rate - becoming shorter at faster rates. It is usually corrected using the cycle length (R-R interval) as shown opposite.
- normal QTc = 0.42 seconds



$$QTc = \frac{QT}{\sqrt{RR}} = \frac{0.71}{\sqrt{1.11}} = 0.67 \text{ seconds}$$

Romano-Ward syndrome is an autosomal dominantly inherited form of long QT interval and there is a risk of recurrent ventricular tachycardia, particularly Torsade de Pointes.

Ventricular premature beats (VPBs)

- 2 ventricular premature beats are also shown in this ECG
- They are
 - broad
 - occur earlier than normal